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Manipal Manual of Surgery

Fourth Edition

with 800 MCQs

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Anitha Shenoy (Nileshwar)

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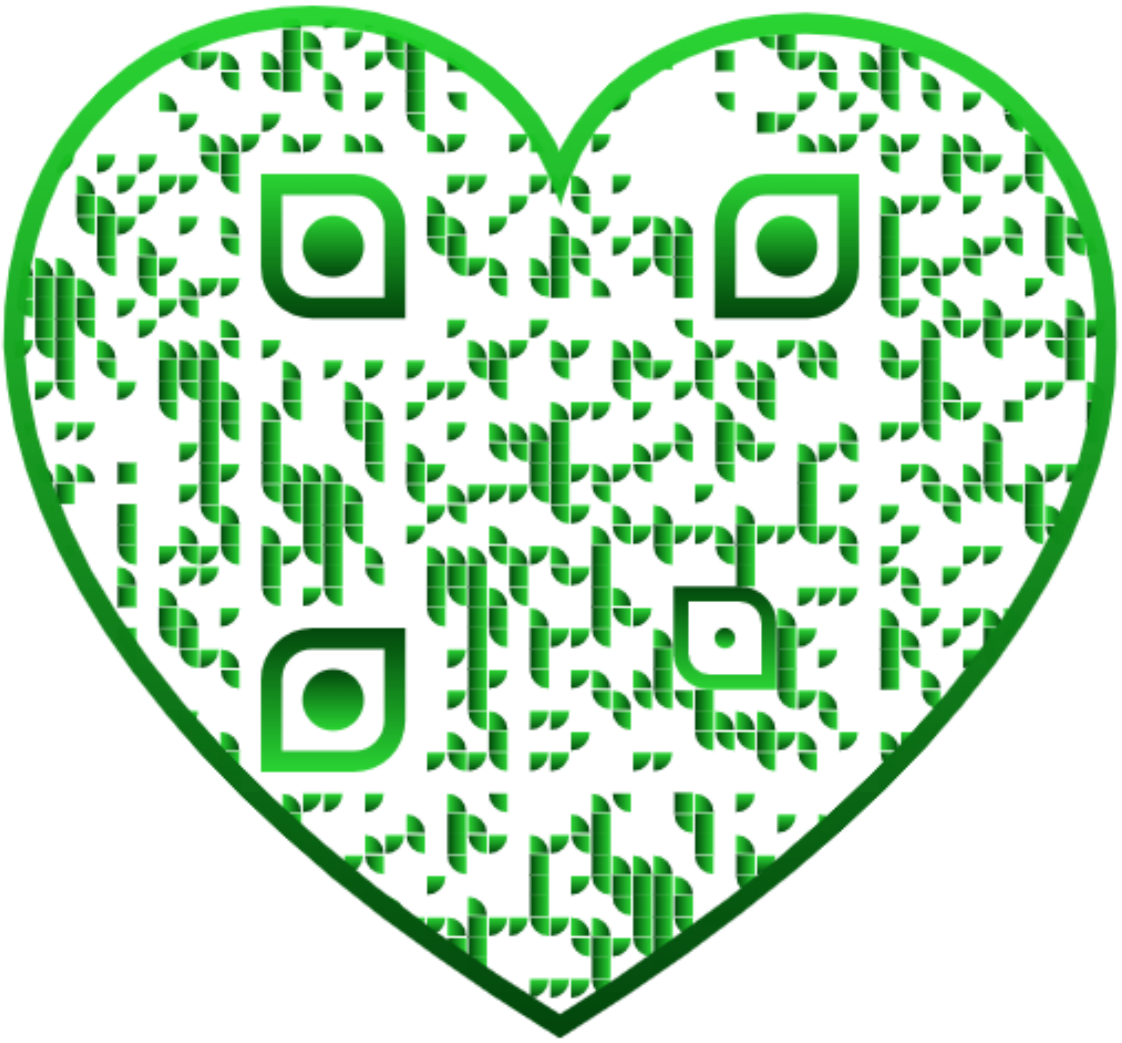

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Section

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1

Wound, Keloid, Hypertrophic Scar and Metabolic Response to Injury

- Types of wounds: Classification
- General principles of management of open wounds
- Components of wound healing
- Factors affecting wound healing
- Compartment syndromes
- Hypertrophic scar and keloid
- Classification of surgical wounds
- Healing of specialised tissues
- Metabolic response to injury
- What is new?/Recent advances

Introduction

Wound is a discontinuity or break in the surface epithelium. A wound is simple when only skin is involved. It is complex when it involves underlying nerves, vessels and tendons.

Types of wounds

A few classifications have been given below.

I A. Closed wounds

- Contusion
- Abrasion
- Haematoma

Contusion: Can be minor soft tissue injury without break in the skin, or major such as when being run over by a vehicle. Generally, it produces discolouration of the skin due to collection of blood underneath.

Abrasion: In this wound, epidermis of the skin is scraped away exposing the dermis. They are painful as dermal nerve endings are exposed. These wounds need cleaning, antibiotics and proper dressings.

Haematoma: This refers to collection of blood usually following injury. It can occur spontaneously in patients who have bleeding tendencies such as **haemophilia**. Depending upon the site, it can be subcutaneous, intramuscular or even subperiosteal. A knee joint haematoma may need to be aspirated followed by application of compression bandage. Small haematomas get absorbed. If not, they can get infected.

B. Open wounds

- Incised
- Lacerated
- Penetrating
- Crushed

Incised wounds: They are caused by sharp objects such as knife, blade, glass, etc. This type of wound has a sharp edge and is less contaminated. Primary suturing is ideal for such wounds, as it gives a neat and clean scar.

Lacerated wounds: They are caused by blunt injury such as fall on a stone or due to road traffic accidents (RTA). Edges are jagged. The injury may involve only skin and subcutaneous tissue or sometimes deeper structures also. Due to the blunt nature of the object, there is crushing of the tissue which may result in haematoma, bruising or even necrosis of the tissue. These wounds are treated by wound excision and primary suturing provided they are treated within six hours of injury.

Penetrating wounds: They are not uncommon. Stab injuries of abdomen are very notorious. It may look like an innocent injury with a small, 1 or 2 cm long cut but internal organs such as intestines, liver, spleen or mesenteric blood vessels may have been damaged. All penetrating wounds of the abdomen should be admitted and observed for at least 24 hours. Layer by layer exploration and repair, though recommended, may not be possible at times due to oblique track of the wound.

Crushed or contused wounds: They are caused by blunt trauma due to run over by vehicle, wall collapse, earthquakes

or industrial accidents. These wounds are dangerous as they may cause severe haemorrhage, death of the tissues and crushing of blood vessels. These patients are more prone for gas gangrene, tetanus, etc. Adequate treatment involves good debridement and removal of all dead and necrotic tissues.

II Tidy and untidy wounds

A. Tidy wounds: Incised, clean, healthy tissue and seldom associated with tissue loss (Key Box 1.1).

B. Untidy wounds: Crushed or avulsed, contaminated, devitalised tissues and often with tissue loss.

KEY BOX 1.1

REPAIR IN TIDY WOUND

- Nerves: Fascicular repair under magnification (loupe or microscope) using 8–0 or 10–0 monofilament nylon
- Artery: To be repaired by using 6–0 prolene
- Tendon repaired by monofilament nonabsorbable suture (polypropylene 3–0 or 4–0)
- Skin loss: Skin flap/skin graft

III Acute wound and chronic wound

A. Acute wound: Stab wounds, following RTA and blast injuries.

B. Chronic wound: Leg ulcers, pressure sores.

General principles of management of open wounds (Fig. 1.1)

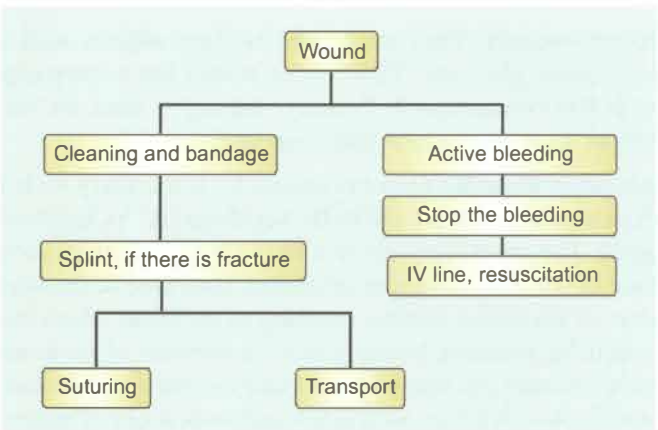


Fig. 1.1: Wound management

- Admission or observation in the hospital.
- Monitoring of temperature, pulse and respiration.
- Systemic antibiotics depending upon the contamination of wound.
- Injection tetanus toxoid for prophylaxis against tetanus.
- Treatment of the wound in the form of cleaning, dressing or suturing.

Healing of the wound

Healing by primary intention occurs in a clean incised wound such as a surgical incision wherein there is only a potential space between the edges. It produces a clean, neat, thin scar.

Healing by secondary intention refers to a wound which is infected, discharging pus or wound with skin loss. Such wounds heal with an ugly scar.

COMPONENTS OF WOUND HEALING (Table 1.1)

I. Inflammatory phase (lag phase)

- Injury results in the release of mediators of inflammation, mainly histamine from platelets, mast cells and granulocytes. This results in increased capillary permeability.
- Later kinins and prostaglandins act and they play a chemotactic role for white cells and fibroblasts.
- In the first **48 hours**, polymorphonuclear (PMN) leukocytes dominate. They play the role of scavengers by removing the dead and necrotic tissue (Figs 1.2 to 1.6).

II. Proliferative phase (collagen phase)

- Between 3rd and 5th days, polymorphonuclear leukocytes diminish in number but monocytes increase. They are the specialised scavengers.
- By 5th or 6th day, **fibroblasts** appear, proliferate and eventually give rise to a protocollagen which is converted

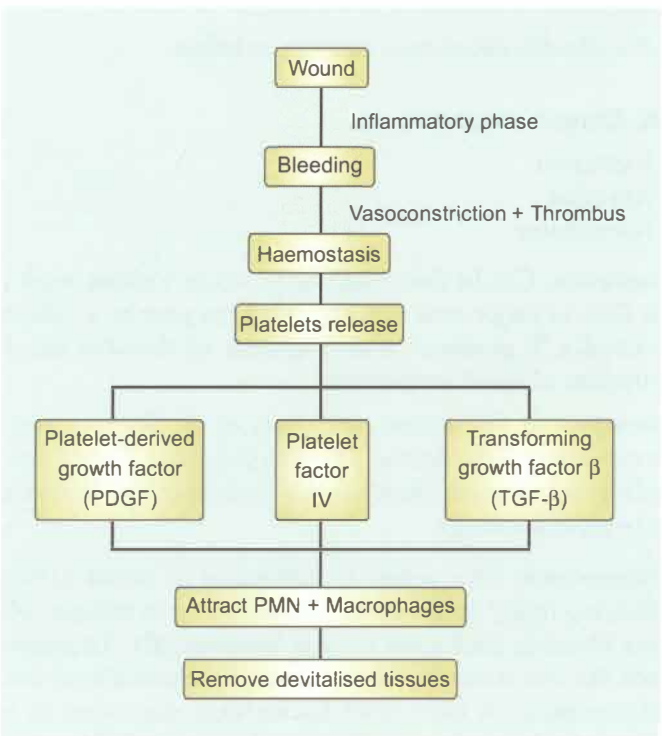


Fig. 1.2: Inflammatory and proliferative phases

into collagen in the presence of an enzyme, protocollagen hydroxylase. O_2, ferrous ions and ascorbic acid are necessary for this step.

- Fibroplasia along with capillary budding gives rise to granulation tissue.

Protocollagen $\xrightarrow[\text{hydroxylation}]{\text{Protocollag enhydroxylase}}$ Collagen

- Secretion of ground substance—mucopolysaccharides by fibroblasts takes place. These are called proteoglycans. They help in binding of collagen fibres. Thus, wound is **Fibre + Gel + Fluid system** (resembles Iron Rods + Cement + Water used for concrete slab).
- **Epithelialisation** occurs mainly from the edges of the wound by a process of cell migration and cell multiplication. This is mainly brought about by **marginal basal cells**. Thus, within 48 hours, the entire wound is re-epithelialised. When there is a wound with skin loss, skin appendages also help in epithelialisation. Slowly, surface cells get keratinised.

III. Remodelling phase (maturation)

It starts after 4 days and is usually completed by 14 days. It is brought about by **specialised fibroblasts**. Because of their contractile elements, they are called **myofibroblasts**. It is the nature's way of reducing the size of defect, thereby helping the wound healing. Wound contraction readily occurs when there is loose skin as in back and gluteal region. Skin contraction is greatly reduced when it occurs over tibia (skin) or malleolar surface. **Corticosteroids, irradiation, chemotherapy delay wound contraction.**

Connective tissue formation: Formation of granulation tissue is the most important and fundamental step in wound healing. (It can be compared to concrete slab laying.)

IV. Phase of scar formation

Following changes take place during scar formation

- Fibroplasia and laying of collagen is increased
- Vascularity becomes less (devascularisation)
- Epithelialisation continues
- Ingrowth of lymphatics and nerve fibres takes place
- Remodelling of collagen takes place with cicatrisation, resulting in a scar.

Complications of wound healing

1. **Infection:** It is the most important complication which is responsible for delay in wound healing. Majority of bacteria are endogenous. Depending upon pus/culture sensitivity report, appropriate antibiotics are given.
2. **Ugly scar:** It is the result of infections
3. **Keloid** and hypertrophic scar (*see* page 10)
4. **Incisional hernia** and wound dehiscence
5. **Pigmentation** of the skin
6. **Marjolin's ulcer** (*see* page 145).

WOUND CLOSURE OR WOUND SUTURING

1. Primary suturing: Suturing the wound within a few hours following an injury (six hours is ideal) is called primary suturing. ***Primary suturing can be done provided:***

Table 1.1 Wound healing

Day 0–1	0–2 days	48–96 hours	5–7 days
• Tissue loss ↓	• Polymorphonuclear cells (PMN) ↓	Macrophages ↓	Fibroblasts appear ↓ — Granulation tissue
• Exposure of extracellular matrix to platelets ↓	• Remove dead and necrotic tissue ↓	Phagocytosis	Bridge the transition of inflammatory to proliferative phase
• Platelet aggregation ↓	• Epithelialisation		• Role of T-lymphocytes is not clear
• Inflammatory mediators			• Wound contraction takes place

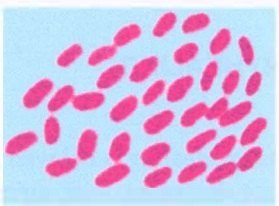


Fig. 1.3: Platelets

Fig. 1.4: Polymorph

Fig. 1.5: Macrophage

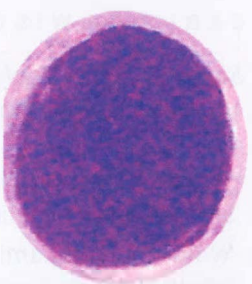


Fig. 1.6: Lymphocytes

- It is an incised or cut wound with a sharp object such as knife or razor blade.
- Minimal injury to structures on either side.
- There should not be any infection. If a wound is sutured in the presence of infection, the suture material is eaten away (digested) by organisms which results in gaping of the wound.

***Precautions to be taken** while doing primary suturing:*

- Foreign body, if present in the deeper aspect of the wound, should be removed.
- Associated injury to blood vessels, nerves or tendons should be recognised and repaired.
- Wound on the abdomen may have associated visceral injuries.
- Prevention of tetanus by administering tetanus toxoid 0.5 ml intramuscularly.

2. Wound excision and primary suturing of skin: This is indicated when:

- Wound edges are jagged.
- Wound is contaminated with organisms or foreign body.
- Tissues are crushed and devitalised.
- In such situations, wound is explored and devitalised tissues and foreign body, if present, are removed. The wound is irrigated with antiseptic agents. Thus, lacerated wound is converted into an incised wound and then sutured.

Precautions to be taken are:

- It should be done within 6 hours.
- Tetanus and gas gangrene prophylaxis.
- Repair of tendons and nerves can be done at a later date, if contamination is excessive.

3. Wound excision and delayed primary suturing: This is indicated in lacerated wounds with major crush injuries. Primary suturing within 6 hours is not done in these wounds because of:

- Gross oedema of the part
- Increased tissue tension
- Haematoma
- Contamination with bacteria
- In such situations, excision of all dead tissues is done.

PEARLS OF WISDOM

Saline is increasingly being used to wash the wound as H_2O_2 and betadine may cause more damage.

- Wound is irrigated with saline and left open without suturing and dressing is applied.
- Wound is re-examined 4–6 days later. If there is no infection, or no nonviable tissues, wound is sutured. This two-stage procedure is called **delayed primary suturing**.

Fig. 1.7: Wound with skin loss

Wound with skin loss (Fig. 1.7): It can follow surgical procedures or accidents, etc.

Principles of debridement

- Ideally done under general anaesthesia
- Assess the extent of injury/loss of tissues
- Control bleeding
- Excision of devitalised tissue better done with scissors
- Good saline wash/irrigation is better than betadine/hydrogen peroxide wash.

PEARLS OF WISDOM

Aim is to convert an untidy wound into a tidy wound.

Complications of skin loss

- Secondary infection of the wound.
- The underlying structures such as tendons and nerves are in danger.
- Diabetic patients can develop septicaemia.
- Deformity and disability can occur at a later date.

Hence, skin grafting should be done as soon as possible.

4. Secondary suturing: After operations, sutures may give way because of severe infection with persistent discharge of pus. In such cases 7–14 days later, after controlling infection, the skin is freed from the edge of the wound and the granulation tissue and skin are approximated. This type of suturing is called as secondary suturing.

FACTORS AFFECTING WOUND HEALING

General factors

1. **Age:** In children, wounds heal faster. Healing is delayed in old age. Dermal collagen content decreases with aging. Also, collagen fibres show distorted architecture and organisation.

2. **Debilitation** results in malnutrition. Wound healing is delayed probably because of vitamin C deficiency. Following injury, vitamin C deficiency can occur after 3–4 weeks. Vitamin C is necessary for the synthesis and maintenance of collagen. **Zinc deficiency is known to delay the healing of pilonidal sinus**. Zinc deficiency is rare—it occurs in large burns, severe polytrauma and hepatic cirrhosis.
3. **In diabetic patients**, wound healing is delayed because of several factors such as **microangiopathy, atherosclerosis, decreased phagocytic activity**, proliferation of bacteria due to high blood sugar, etc. (Key Box 1.2)

   Also poor immune response is seen in diabetic patients.

KEY BOX 1.2

DIABETES AND WOUND HEALING FACTORS

Tissue hypoxia due to atherosclerosis
Thickened basement membrane→↓ tissue perfusion
Trauma—repetitive due to neuropathy
Tissue metabolism is increased—relative hypoxia
Total failure of defence mechanism

Observe **5 T**s

4. **Jaundiced** and **uraemic** patients have poor wound healing because fibroblastic repair is delayed.
5. **Cytotoxic** drugs such as doxorubicin and **malignancy delay healing** (Key Box 1.3).

KEY BOX 1.3

CHEMOTHERAPEUTIC DRUGS

- Decrease mesenchymal cell proliferation
- Reduce number of platelets
- Reduce inflammatory cells
- Reduce growth factors
- Decrease wound breaking strength

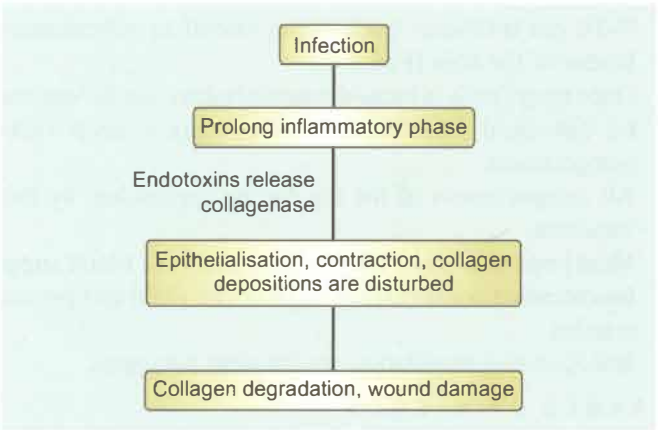
6. **Generalised infection:** Pus in some part of body delays wound healing.
7. **Corticosteroids** given early may delay wound healing because of their anti-inflammatory activity. Once healing is established, they do not interfere.
8. **Malnutrition:** Definitely results in delayed wound healing including intestinal anastomotic leakage and wound dehiscence.

PEARLS OF WISDOM

Albumin levels must be less than 2 g /dL to have an effect on wound healing.

Local factors

1. **Poor blood supply:** Wound over the knee and shin of tibia heals very slowly but wound on the face heals fast.
2. **Local infection:** Organisms eat away the suture material, destroy granulation tissue and causes slough and purulent discharge. If the bacterial count exceeds 10^5 organisms/mg tissue or if any β-hemolytic streptococci are present, **wound does not heal**. Collagen synthesis is reduced and collagenolysis is increased. Antibiotics should be given immediately or within 2 hours to prevent infection.



3. **Haematoma** precipitates infection.
4. **Faulty technique** of wound closure.
5. **Tension** while suturing.
6. **Hypoxia:** Killing property of macrophages and production of fibroblasts can decrease due to hypoxia. If contamination occurs, oxygen level in tissue decreases.
   - Collagen synthesis is affected in cases of hypoxia. In anaemic patients, wound healing is delayed because of decreased angiogenesis and decreased collagen production.
   - Smoking causes vasoconstriction and elevated carbon monoxide levels.
7. **Ionising radiation:** It causes endothelial cell injury with endarteritis and results in atrophy and fibrosis.
   - Typically occur in closed lower limb injuries.
   - Following injuries, inflammatory reaction results in gross oedema of the region.
   - These are the tight undividing compartments in the leg containing nerves and vessels.

PEARLS OF WISDOM

The most prone site is the anterior compartment. Paraesthesia or numbness between 1st and 2nd toes occurs due to pressure on the deep peroneal nerve and is diagnostic.

Clinical features

- Severe pain in the leg
- Sensory disturbances (Fig. 1.8)
- Colour changes—absence of pulses is a late sign.

Measuring compartmental pressure

- It is measured using a catheter placed in the muscle compartment and a pressure transducer.
- **Compartmental pressure greater than 30 mmHg** is an indication for **urgent fasciotomy**.

Treatment—Fasciotomy

- 8–10 cm incisions, each being lateral to subcutaneous border of the tibia (Fig. 1.9).
- Once deep fascia is incised, muscle bulges out. Soleus must be detached from tibia to decompress deep flexor compartment.
- All compartments of the leg can be approached by these incisions.
- Most important motto should be to **preserve blood supply by releasing compression** on posterior tibial and peroneal arteries.
- Infection and amputation are frequent outcomes.

PEARLS OF WISDOM

The danger lies in the delay, not in the simple fasciotomy.

In crush injuries who present late to the hospital, it is safer to do amputation because by doing fasciotomy sudden release of myoglobin from dead muscle may cause systemic myoglobinuria, glomerular blockage and renal failure.

HYPERTROPHIC SCAR AND KELOID

- As the name suggests, there is hypertrophy of mature fibroblasts in hypertrophic scar. Blood vessels are minimal in this condition. However, in keloid, proliferation of immature fibroblasts with immature blood vessels are found. These two conditions represent variations in the normal process of wound healing (Table 1.2).
- Keloid is **very common in blacks** and least common in Caucasians (Key Boxes 1.4 and 1.5).

KEY BOX 1.4

TYPES OF SCAR

- Atrophic
- Hypertrophic
- Keloid

KEY BOX 1.5

AETIOLOGY OF KELOID

- **K**ey factors: Surgery, burns, vaccinations
- **E**levated levels of growth factor (more of type 'B' collagen)
- **L**aceration or abrasion
- **O**ver the sternum (incision)
- **I**nheritance and injection
- **D**eep pigmented skin

Remember as **KELOID**

- Keloid is **not a true tumour** but has a marked tendency for **local recurrence** after excision.
- Keloid takes the shape of a butterfly over the sternum. It is the commonest site for a keloid. It is extremely difficult to treat the keloid over the sternum. We had one patient who underwent wide excision and grafting 6 times for a sternal keloid. Jaw, vaccination are common sites (Figs 1.10 to to 1.13 and Key Box 1.6).

What is a contracture?

When scar crosses 'joints or flexion creases, a tight 'web' may form which is referred to as a contracture (*see* Burns Chapter for more details).

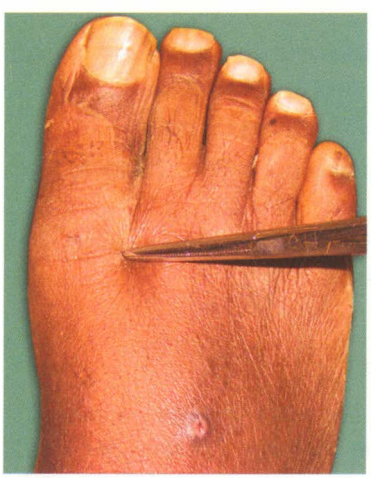


Fig. 1.8: Paraesthesia between first and second toes

Fig. 1.9: Fasciotomy

Fig. 1.10: Keloid over the sternum

Table 1.2 Comparison of hypertrophic scar and keloid (Figs 1.11 to 1.13)

Hypertrophic scar	Keloid
General features	
• It occurs from a prolonged inflammatory phase of wound healing. It never gets worse after 6 months.	• It continues to get worse even after 1 year and up to a few years.
• Itching is not usually present. If present, it is not severe.	• Severe itching is present
• Nontender	• Margin is tender
• Not vascular	• Vascular, red, erythematous (immature blood vessels)
• Does not extend beyond the boundary of the original incision or wound. It rises above skin.	• Extends to normal tissues, has **claw-like** process. Hence the name.
Precipitating factors	
• Scar crossing normal skin creases	• Black race
• Over sternum, over joints	• Tuberculosis patients
• Young persons	• Incision over the sternum, ear lobe
	• Equal in both sexes
	• Hereditary and familial
	• Vaccination sites, injection sites, incision sites, piercing sites
Natural history	
• May become small	• Does not become small
Complications	
• Do not occur	• Ulceration, infection
Treatment	
• It is often not necessary	• It is difficult. Injection of steroid preparation such as triamcinolone acetate (Kanacort) has been found to be extremely useful. It flattens the keloid. **Intrakeloidal excision** and skin grafting is to be tried last. Recurrence is common. Care should be taken **not to extend** the incision on to the normal surrounding tissues.
• Stocking, armlets, gloves	• Silicone application
• Elastic bandage may help	• Topical retinoids
• Excision can be done	
• Silicone application	
• Topical retinoids	

PEARLS OF WISDOM

Any form of excision has high chance of recurrence

Fig. 1.11: Keloid over the jaw

Fig. 1.12: Recurrent keloid over the sternum. Excision attempted three times

KEY BOX 1.6

KELOID SITES

High chances	*Least chances*
Skin of ear lobe	Eyelid
Presternal	Genitalia
Deltoid	Palm, sole
Upper back	Across joint

PEARLS OF WISDOM

Collagen bundles are virtually absent in keloid. It is extremely difficult to treat a keloid.

Fig. 1.13: Keloid over vaccination site

SURGICAL WOUNDS

Surgical wounds may be classified depending upon the nature of the wound, whether clean or contaminated (Table 1.3 and Figs 1.14 to 1.17).

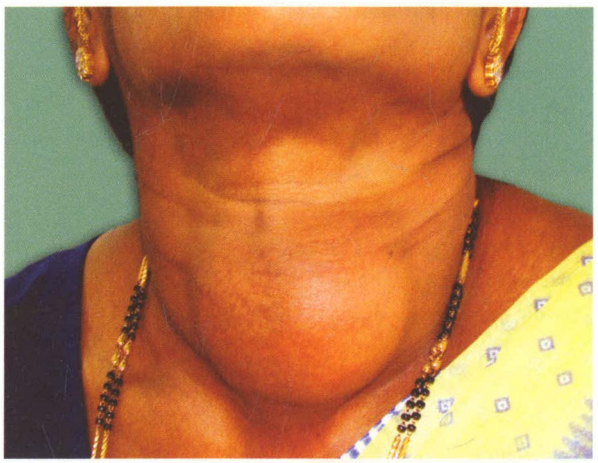


Fig. 1.14: Excision of neck swelling—clean

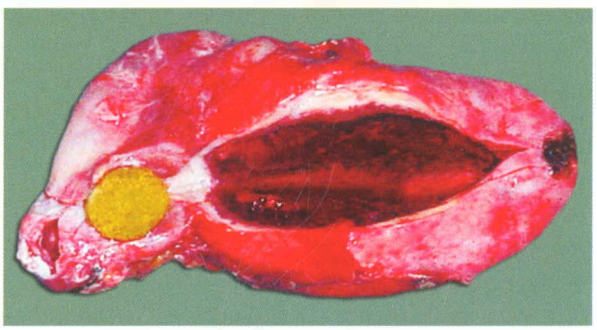


Fig. 1.15: Cholecystectomy—clean contaminated

Table 1.3 Classification of surgical wounds

Wounds class	Definition	Examples of typical procedures	Wound infection rate (%)	Usual organism
Clean (Fig. 1.14)	• Nontraumatic • Elective surgery • Gastrointestinal, respiratory or genitourinary tract not entered.	• Mastectomy • Vascular procedures • Thyroidectomy	2	*Staphylococcus aureus*
Clean contaminated (Fig. 1.15)	• Respiratory, genitourinary or gastrointestinal tracts entered but minimal contamination	• Gastrectomy • Hysterectomy • Cholecystectomy	< 10	Related to viscus entered
Contaminated (Fig. 1.16)	• Open, fresh, traumatic wounds • Uncontrolled spillage from an unprepared viscus • Minor break in sterile technique	• Ruptured appendix • Resection of unprepared bowel	20	Depends on underlying cause
Dirty (Fig. 1.17)	• Open, traumatic dirty wounds • Traumatic perforated viscus • Pus in the operative field	• Resection of gangrene	30–70	Depends on underlying cause

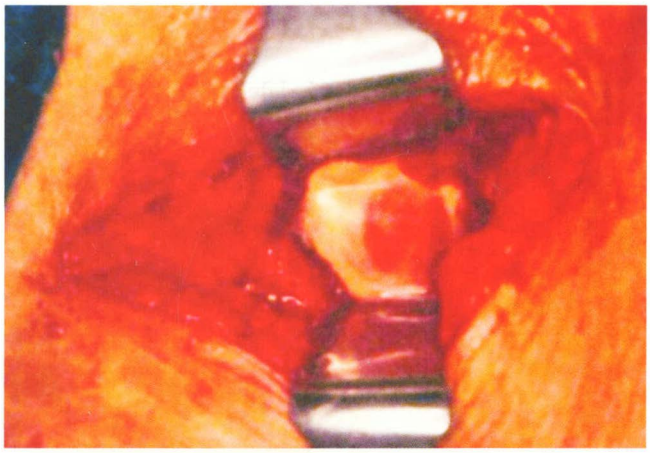


Fig. 1.16: Appendicular abscess—contaminated

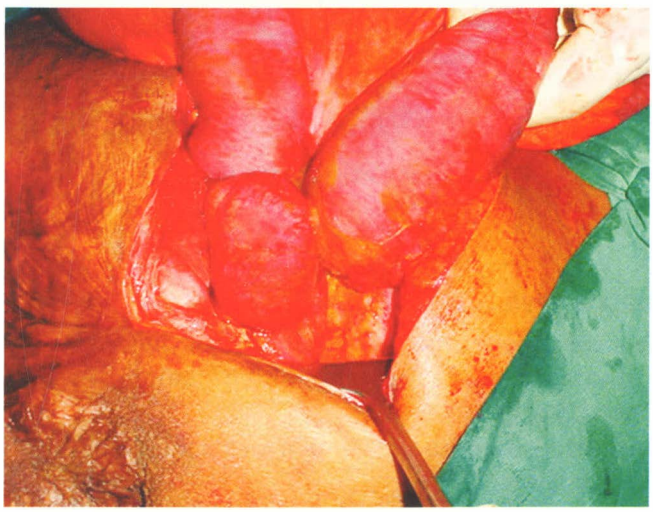


Fig. 1.17: Peritonitis—dirty

MISCELLANEOUS

HEALING OF SPECIALISED TISSUES (Key Box 1.7)

KEY BOX 1.7

HEALING OF SPECIALISED TISSUES ONCE DESTROYED

• Nerve cells of brain and spinal cord	• Cannot be replaced by proliferation of other nerve cells
• Peripheral nerves	• Regenerative capacity is present
• Stomach and intestines	• Healing is good after anastomosis, rarely leaks
• Colon and oesophagus	• Healing is precarious, chances of leakage are high.
• Wounds on the face	• Healing is excellent due to good vascularity
• Muscles	• Can heal completely or may be replaced by fibrosis
• Bone	• Rapid proliferation of osteoblasts

METABOLIC RESPONSE TO INJURY

Sir Davis Cuthbertson (1930) proposed that human beings respond to any injury in two distinct phases, namely the 'ebb phase' and the 'flow phase'. The 'ebb phase' begins immediately following surgery and lasts for 24 to 48 hours. This phase aims to conserve circulating fluid volume and energy. The 'ebb phase' is characterised by hypovolaemia, hypothermia, reduced basal metabolic rate (BMR), reduced cardiac output and lactic acidosis. Renin-angiotensin cascade is an important component of the 'ebb phase'. The hormones responsible are adrenal hormones, i.e. catecholamines, cortisol and aldosterone. The 'flow phase' follows the 'ebb phase'. This phase is concerned with recovery and repair following the initial injury. The 'flow phase' is subdivided into catabolic phase and the anabolic phase in the order of evolution.The catabolic phase is associated with increased levels of catecholamines, cortisol, insulin and glucagon. Following issues are important aspects of metabolic response to stress.

1. Energy expenditure

During stress, energy expenditure **increases by 15–25%.** Causes for such an increase are:

- Central thermodysregulation caused by pro-inflammatory cytokines
- Increased sympathetic activity
- Increased lactate production in ischaemic areas which in turn is metabolised by energy consuming **Cori cycle**
- Increased cardiac output
- Increased protein turnover

2. Changes in skeletal muscle

During catabolic phase of stress response, there will be muscle wasting. Causes for such muscle wasting include:

- Increase in muscle protein degradation
- Decrease in synthesis of muscle protein.

Such protein catabolism results in release of amino acids, **mainly alanine and glutamine,** which are used by liver and immune system, for the synthesis of proteins such as acute phase proteins and cytokines. Although all types of muscles contain proteins, proteins from skeletal and smooth muscles are preferentially catabolised (in that order), when compared to cardiac muscles. **The main pathways which govern protein catabolism in skeletal muscles are:**

- ATP dependent ubiquitin proteasome pathway
- Lysosomal cathepsin
- Calcium calpain pathway.

Exaggerated skeletal muscle catabolism results in asthenia, increased fatigability, poor quality of life, increased morbidity and mortality. The basic physiological defect in such condition will be impaired excitation-contraction coupling at the level of sarcolemma and sarcoplasmic reticulum. This condition has been termed as 'critical illness myopathy'.

Changes in liver protein metabolism

Normally liver synthesises two types of proteins:

1. Structural proteins
2. Export proteins, e.g. albumin.

During stress, liver synthesises

1. Positive acute phase reactants, e.g. fibrinogen and C-reactive proteins (CRP).
2. Negative acute phase reactants, e.g. albumin.

Albumin and its clinical significance

Albumin is the major protein synthesised by liver. Hypoalbuminemia is commonly associated with malnutrition and severe stress/sepsis. It is tempting to presume that fall in albumin level during stress is due to diversion of synthetic activity of liver towards synthesis of positive acute phase reactants. This might not be true. Serum level of albumin is governed by rate of synthesis and transcapillary escape rate. During stress, transcapillary escape rate of albumin increases by about 3 times. This is because of increased microvascular permeability, which results in hypoalbuminemia during stress/sepsis.

RESPONSE TO INJURY

Introduction

The response of body (milieu interieur) to injury include physiological and immunological changes. Such response is a dynamic event, and depends upon the severity of the injury. The response of body to injury can be considered under the following headings:

- Neuroendocrine response
- Immunological response
- Metabolic response

Neuroendocrine response (Fig. 1.18)

One of the earliest pathways to get activated following injury is the "neuroendocrine pathway". The neuroendocrine pathway starts with nociceptive receptors involves spinal cord, thalamus, hypothalamus and pituitary. Stimulation of hypothalamus is a key event in the pathway and results in:

- Release of corticotrophin releasing hormone (CRH), which triggers the release of ACTH from the anterior pituitary. ACTH acts on adrenals to increase the secretion of cortisol.
- Activation of sympathetic nervous system, which in turn causes release of adrenaline and glucagon.
- Increased release of growth hormone and glucagon.

To summarise, the levels of ACTH, cortisol, growth hormone, adrenaline and glucagon levels are increased following injury mainly because of activation of neuroendocrine pathway.

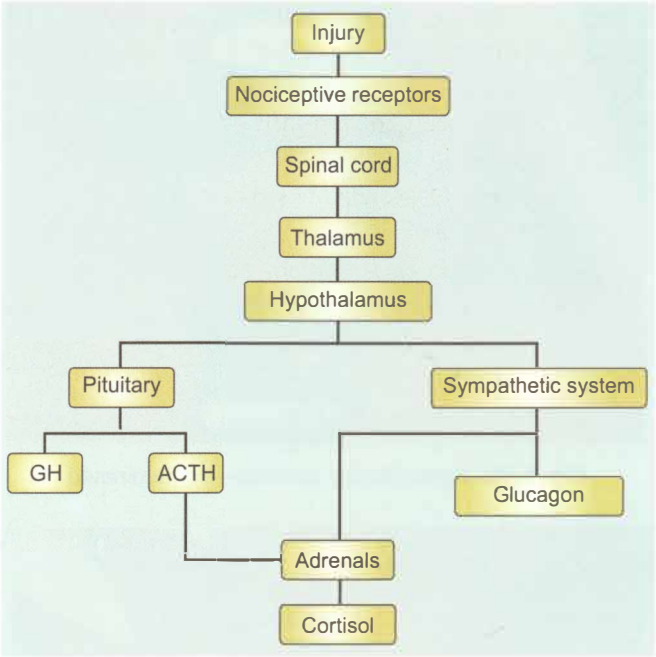


Fig. 1.18: Neuroendocrine response

Immune response (Fig. 1.19)

The immune system of the body may be subdivided as:

- **Innate immune system** which mainly includes macrophages
- **Adaptive immune system** which includes T and B lymphocytes

Immune responses are mediated by protein signalling compounds called "cytokines". The immune response to injury includes pro-inflammatory response and anti-inflammatory response. Depending upon the type of immune response mediated by the cytokines, they can be designated as pro-

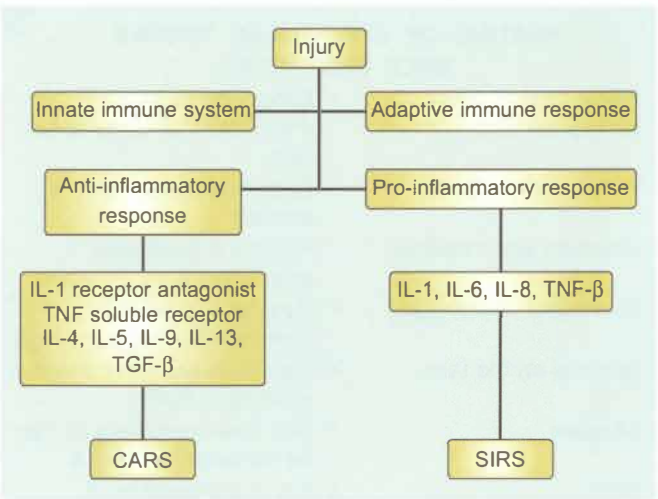


Fig. 1.19: Immune response

inflammatory cytokines and anti-inflammatory cytokines. The initial response to injury is pro-inflammatory and results are such as pyrexia, proteolysis, propagation, productions.

- Pyrexia, due to action of pro-inflammatory cytokines on hypothalamus
- Augmentation of hypothalamic stress response
- Proteolysis in skeletal muscle
- Production of acute-phase proteins in liver

KEY BOX 1.8

POINTS TO REMEMBER

Hormones whose levels rise following injury:

- ACTH
- GH
- Glucagon
- Cortisol
- Adrenaline

Hormones whose levels fall following injury:

- Insulin
- Thyroid hormones
- Testosterone

Pro-inflammatory cytokines:

- IL–1
- IL–6
- IL–8
- TNF α

Anti-inflammatory cytokines:

- IL–4
- IL–5
- IL–9
- IL–13
- TNF β

The pro-inflammatory cytokines are interleukin-1(IL–1), tumour necrotic factor-α, interleukin-6 (IL–6), and interleukin-8 (IL–8). Following pro-inflammatory response endogenous pro-inflammatory cytokine antagonists like IL-1 receptor antagonist and TNF-soluble receptors are released into circulation which serves to counteract pro-inflammatory cytokines. Further adaptation of body results in development of Th 2-type counter inflammatory response. Such counter inflammatory response is mediated by IL–4, IL–5, IL–9, IL–13, and transforming growth factor-β. There is a delicate balance between pro-inflammatory and anti-inflammatory immune response. Unopposed pro-inflammatory response leads to "Systemic immune response syndrome". On the other hand, exaggerated anti-inflammatory response results in "Compensatory anti-inflammatory response syndrome (CARS)" or "Counter inflammatory response syndrome".

Summary of response to injury has been given in Key Box 1.8.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- Wound healing is edited.
- Compartment syndrome is added with an approach to arrive at a clinical diagnosis.
- Debridement principles are added.
- Please remember usage of H_2O_2 and Eusol have diminished as they may cause more tissue damage.
- Role of collagen is added.
- Metabolic response to injury has been added.

MULTIPLE CHOICE QUESTIONS

1. Platelet aggregation occurs in which phase of the wound healing?

A. Inflammatory phase
B. Proliferative phase
C. Remodelling phase
D. Scar formation phase

2. Chronic scar can change into:

A. Martorell's ulcer
B. Marjolin's ulcer
C. Meleney's ulcer
D. Malignant melanoma

3. The ideal solution to wash the wound is:

A. Saline
B. Betadine
C. Hydrogen proxide
D. Eusol

4. Following facts are true in wound healing *except*:

A. Vitamin C is necessary for synthesis of collagen
B. Diabetic patients have decreased phagocytic activity
C. Once healing is established, corticosteroids can be given
D. Poor wound healing in jaundiced patients is due to poor functioning of macrophages

5. Following are true in compartment syndromes *except:*

A. Usually occur in the lower limbs
B. Most prone site is posterior compartment
C. Diagnostic feature is paraesthesia between 1st an 2nd toe
D. Compartment pressure greater than 30 mm of H is an indication for urgent fasciotomy

6. Following are true for keloid *except:*

A. It extends into normal skin
B. It continues to become worse even after several years
C. It does not give rise to itching
D. It recurs often after excision

7. Following are true for hypertrophic scar *except:*

A. It does not extend into normal skin
B. It does give rise to itching
C. It becomes worse even after 2 years
D. It is more common in blacks

8. Following are true for keloid *except:*

A. Sternum is one of the common sites for keloid
B. Collagen bundles are present in keloid
C. Intrakeloidal injections can be used to treat
D. Deep pigmented skin is more vulnerable fo keloid

9. Which hormone level does not rise during injury?

A. ACTH
B. Adrenaline
C. Glucagon
D. Thyroxine

10. Albumin level decreases during stress because of:

A. Decrease production
B. Decrease intake
C. Liver failure
D. Increased microvascular permeability

ANSWERS

1 A 2 B 3 A 4 D 5 B 6 C 7 C 8 B 9 D 10 A

2

Acute Infections, Sinus, Fistula and Surgical Site Infection

- Cellulitis
- Ludwig's angina
- Lymphangitis
- Abscess
- Cervical tuberculous lymphadenitis
- Boil, carbuncle
- Erysipelas
- Chronic abscess
- Necrotising fasciitis
- Acute pyomyositis
- Surgical site infections
- Asepsis and antisepsis
- Sinus and fistula
- What is new? / Recent advances

Introduction

Since the time surgery has evolved as a speciality, infection and haemorrhage have been recognised as two well-known enemies of surgeons. Over a period of time, many newer antibiotics have come into existence. However, infection still dominates and it is one of the major causes of mortality and morbidity in a patient who has a 'benign disease'. This is the sad part of consequence of disease. Hence, it is important to diagnose and treat infections effectively as early as possible.

CELLULITIS

Cellulitis is a spreading subcutaneous inflammation caused by *haemolytic Streptococcus*. ***Streptococci produce hyaluronidase and streptokinase.*** Net result is that the inflammatory exudate spreads in the subcutaneous and fascial planes resulting in a gross swelling of the affected part. Wherever there is loose subcutaneous tissue, as in scrotum or loose connective and interstitial tissue as in face and forearm, it spreads fast.

Sources of infection

- Injuries—minor or major
- Graze or scratch
- Snake bite, scorpion bite, etc.

Precipitating factors

- Diabetes
- Low resistance of an individual

Common sites

- Lower limbs
- Face
- Scrotum

Clinical features

The affected part shows evidence of inflammation such as redness and itching followed by diffuse swelling. Skin is stretched and shiny. Pain, fever, toxaemia follow later. It is differentiated from an abscess by features mentioned in Key Box 2.1.

In untreated cases, suppuration, sloughing and gangrene can occur (Fig. 2.1).

Fig. 2.1: Cellulitis of the foot with abscess

KEY BOX 2.1

CELLULITIS	ABSCESS
• No edge (diffuse swelling)	• Well-circumscribed, has an edge
• No limit	• Limit is present
• No pus	• Pus is present
• No fluctuation	• Fluctuation is positive

Treatment

- Bed rest with legs elevated. This reduces oedema of legs.
- Glycerine $MgSO_4$ dressing which reduces oedema of the part by osmotic effect.
- Diabetes mellitus, if present, is treated with injection **insulin** given **subcutaneously**.
- Appropriate antibiotics such as injection crystalline penicillin 10 lakh units, intramuscular (IM) or intravenous (IV), 6th hourly for 5–7 days or cephalosporins have to be given.
- Antisnake venom is given in snake bite cases.

Complications

1. Cellulitis can turn into an **abscess** which needs to be drained.
2. **Necrotising fasciitis:** Certain highly invasive strains of *Streptococcus pyogenes* can cause extensive necrosis of skin, subcutaneous tissues and may result in necrotising fasciitis. It is treated by debridement and skin grafting later (*see* page 17).
3. **Toxaemia and septicaemia:** *Streptococcal toxic shock syndrome* can result if exotoxins are produced by the organisms.
4. Cellulitis can precipitate **ketoacidosis** in a patient who has diabetes mellitus.

LUDWIG'S ANGINA

It refers to cellulitis of submental and submandibular regions combined with inflammatory oedema of the mouth. Virulent streptococcal organisms are responsible for infection surrounding submandibular region. Anaerobes also play a major role (Key Box 2.2).

KEY BOX 2.2

PRECIPITATING FACTORS

Caries tooth
Cancer of the oral cavity
Calculi in the submandibular gland
Chemotherapy
Cachexia
Chronic disease—diabetes

Observe **6 Cs**

Clinical features

- Elderly patient who presents with diffuse swelling in th submandibular and submental region (Brawny oedema).
- **Oedema of the floor of the mouth**, as a result of whicl the tongue is pushed upwards resulting in difficulty i swallowing.
- High grade fever with toxicity.
- **Putrid halitosis** is characteristic of this condition.

Treatment

- Rest and hospitalisation
- Appropriate antibiotics
- Intravenous fluids to correct dehydration and Ryle's tub feeding.
- If it does not respond to conservative treatment, surgica intervention is recommended.

Surgery

Under general anaesthesia, 5–6 cm curved incision is mad below the mandible in the submandibular region over the most prominent part of the swelling. Submandibular gland is mobilised, mylohyoid muscle is divided and the pus is drained. ***Even if pus is not found, the oedematous fluid comes out greatly improving the condition of the patient.*** Wound is closed with loose sutures, after irrigating the cavity with antiseptic agents and a drainage tube is kept in place.

Complications

1. Mediastinitis and septicaemia
2. Oedema of the glottis due to spread of the cellulitis *via* a tunnel occupied by stylohyoid to submucosa of the glottis.

LYMPHANGITIS

- It is also a nonsuppurative, poorly localised infection caused by *streptococci, staphylococci* or *clostridia.*
- It presents as red painful streaks in affected lymphatics.
- **Filarial infection** is one of the common causes of lymphangitis in coastal India (*see* page 112).
- **High grade fever, chills and rigors** and features of systemic inflammatory response syndrome (SIRS) are common. **Tender, painful lymph nodes** in the groin are characteristics of lower limb lymphangitis.

Treatment

- Rest, elevation, $MgSO_4$, local dressing, antibiotics, anti-inflammatory drugs.
- Anti-filarial treatment in appropriate cases.

ABSCESS

An abscess is a localised collection of pus (dead and dying neutrophils plus proteinaceous exudate).

CLASSIFICATION

1. **Pyogenic abscess:** It is the commonest form of an abscess. It can be subcutaneous, deep or can occur within the viscera such as liver or kidney. In this chapter, pyogenic abscess refers to soft tissue abscess.
2. **Pyaemic abscess:** Occurs due to circulation of pyaemic emboli in the blood (pyaemia).
3. **Cold abscess:** Usually refers to tubercular abscess due to involvement of either lymph nodes or spine.

PYOGENIC ABSCESS

It is usually produced by ***Staphylococcal infections***. The organisms enter soft tissues through an external wound, minor or major. It can also be due to haematogenous spread from a distant focus such as tonsillitis or caries tooth. Pyogenic abscess can also be due to cellulitis.

Pathophysiology

- Following an injury, there is inflammation of the part brought about by the organism such as *Staphylococcus*. Pathological events are summarised in Fig. 2.2.
- The end-result is production of pus which is composed of dead leukocytes, bacteria and necrotic tissue. The area around the abscess is encircled by fibrin products and it is infiltrated with leukocytes and bacteria. It is called **pyogenic membrane**.

Symptoms

The patient feels ill and complains of **throbbing pain** at the site. Throbbing pain is indicative of pus and is due to pressure on the nerve endings by the pus. Fever, with or without chills and rigors, can be present.

Signs

1. **Calor**—heat: The affected part is warmer due to local rise in temperature.
2. **Rubor**—redness: It is due to inflammation resulting in hyperaemia.
3. **Dolor**—pain: An abscess is extremely tender.
4. **Tumour**—swelling: It consists of pus. It is tensely cystic with surrounding brawny oedema.
5. **Loss of function:** The function of the part is impaired, due to pain.
6. **Fluctuation:** It can be elicited. However, in a deep-seated abscess it may be negative, as in breast abscess.

Treatment

- Untreated abscess tends to point spontaneously along the area of least resistance to the nearest epithelial surface, e.g. skin, gut, oral cavity. However, deep-seated abscess such as breast abscess may cause much tissue destruction before pointing.
- **Incision and drainage (I&D)** under general anaesthesia. General anaesthesia is preferred because in the presence of infection, local anaesthesia may not act and it is difficult to break all the loculi of an abscess without causing pain.

Procedure

- A **stab** incision is made over the most prominent (pointing) part of the abscess. The pus which comes out is collected and sent for culture and sensitivity. A sinus forceps or a finger is introduced within the abscess cavity and all the loculi are broken down. Fresh oozing of the pus is an indication of completion of the procedure. The abscess cavity is irrigated with saline or mild antiseptic agents such as iodine solution or hydrogen peroxide. Hydrogen peroxide acts by liberating nascent oxygen.

$$H_2O_2 \rightarrow H_2O + [O]$$

- Nascent oxygen bubbles out and thus helps in separating the slough. The cavity, if large, may need to be packed with roller gauze dipped in iodine solution which is

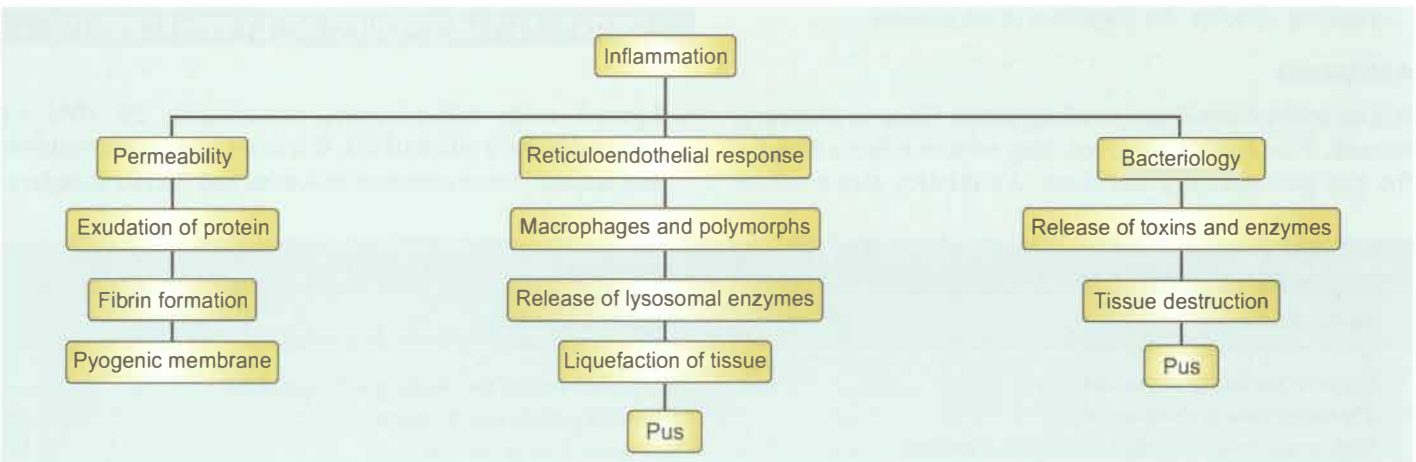


Fig. 2.2: Pathological events during inflammation

removed 1–2 days later. Roller gauze packing prevents the premature closure of the skin, thereby facilitating the healing to take place from the depth of the cavity by granulation tissue formation. With appropriate antibiotics and proper dressings, the wound heals within 5–7 days.

- Antibiotic of choice is cloxacillin for staphylococcal abscess. Dosage: 500 mg, 6th hourly for 5–7 days.
- **Modified Hilton's method** for I&D. This method is followed where an abscess is situated in the vicinity of important anatomical structures such as vessels or nerves (Table 2.1).
- In this method skin and superficial fascia are incised, instead of a stab incision, followed by opening of the abscess by sinus forceps, so as to avoid damage to vital structures such as vessels and nerves.

Table 2.1 Relationship of nerves or vessels with an abscess

Site		Anatomical structure
Axilla	→	Axillary vessels
Neck	→	Subclavian vessels and brachial plexus
Parotid region	→	Facial nerve
Midpalmar space	→	Median nerve

Differential diagnosis

1. **Ruptured aneurysm** can present as subcutaneous abscess with pain, redness and local rise of temperature. There may be leukocytosis also. Ruptured vertebral artery aneurysm in the posterior triangle and popliteal artery aneurysm in the popliteal fossa have been incised, mistaking them for an abscess (Table 2.2).
   Caution: When in doubt, aspirate with a wide bore needle before incising an abscess.
2. **Soft tissue sarcoma** in the thigh can be confused for a deep-seated abscess. However, throbbing pain, high grade fever with chills and rigors and short duration of the swelling clinches the diagnosis of an abscess.

Antibioma

It is an antibiotic-induced swelling (oma). Once an abscess is formed, if antibiotics are given, they seldom effect a cure but the pus gets partially sterilised. Antibiotics also produce fibrosis, resulting in thickening of the abscess wall. Clinically this may result in a **hard lump**.

Sites of antibioma are **breast, thigh, ischiorectal fossa**, etc. Antibioma in the breast may mimic carcinoma of the breast.

PYAEMIC ABSCESS

This is due to pus-producing organisms in the circulation (pyaemia). It is the systemic effect of sepsis. It commonly occurs in diabetics and patients receiving chemotherapy and radiotherapy. Pyaemic abscess is characterised by following features:

- They are multiple
- They are deep-seated
- Tenderness is minimal
- Local rise of temperature is not present.

Hence, it is called **nonreactive abscess** to differentiate it from pyogenic abscess. This is treated by multiple incisions over the abscess site and drainage (like a pyogenic abscess) with antibiotic cover.

COLD ABSCESS

- Even though it is a chronic abscess due to a chronic disease (tuberculosis), for the completeness of the chapter on abscess and for the convenience of reading, it is discussed here.
- Cold abscess means an abscess which has no signs of inflammation. Usually, it is due to tuberculosis, e.g. following tubercular lymphadenitis or due to tuberculosis of spine. However, other chronic diseases such as leprosy, actinomycosis and madura foot also produce abscesses which are 'cold' in nature (Key Box 2.3). In this chapter, cold abscess due to tubercular lymphadenitis in the neck is discussed.

KEY BOX 2.3

COLD ABSCESS—CAUSES

- Tuberculosis (mainly *refers* to TB)
- Actinomycosis
- Leprosy
- Madura foot

CERVICAL TUBERCULOUS LYMPHADENITIS

- Lymph node tuberculosis constitutes 20–40% of extrapulmonary tuberculosis. It is more common in children and women, more common in Asians and Pacific islanders.

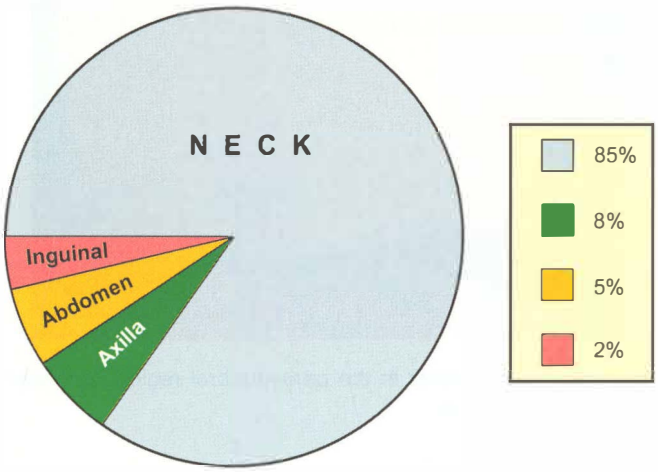
Table 2.2 Differences between acute abscess and ruptured aneurysm

Acute abscess	Ruptured aneurysm
No previous history of the swelling	Previous history of the swelling will be present
Throbbing pain is characteristic	Throbbing pain is usually absent
High grade fever with chills and rigors is present	Low grade fever is present
Extremely tender	Tender

- The disease may be caused by *Mycobacterium tuberculosis*, atypical mycobacteria and *Mycobacterium bovis*..

Aetiopathogenesis: In 80% of the cases, mycobacteria pass through tonsillar crypts and affect tonsillar node or jugulodigastric group of nodes, in the anterior triangle of the neck.

- In 20% of the cases, lymph nodes in the posterior triangle are affected due to involvement of adenoids.
- Rarely, infection can spread from tuberculosis of apex of lung. Organisms directly penetrate Sibson's fascia (suprapleural membrane) and can cause enlargement of supraclavicular node.
- Other lymph nodes in the neck such as preauricular, submandibular can also be affected.
- In general, the incidences of lymphadenopathy at various sites are as follows:



Incidence of lymph node tuberculosis

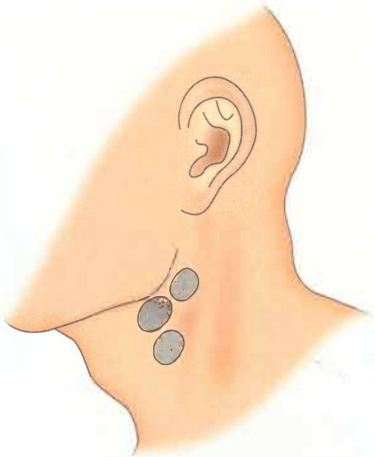


Fig. 2.3: TB lymphadenitis—discrete nodes

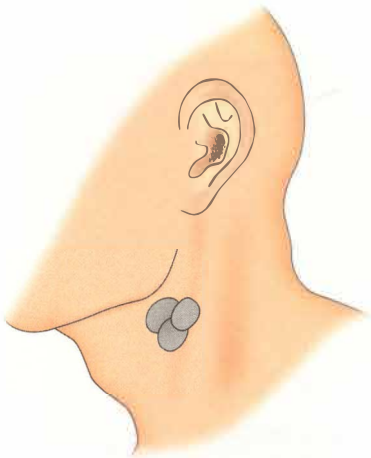


Fig. 2.4: TB lymphadenitis—matted nodes

Clinical features

- Tuberculous lymphadenitis presents as a gradually increasing painless swelling of one or more lymph nodes of a few weeks to a few months duration. Multiple sites may be involved.
- Systemic symptoms such as fever, weight loss, fatigue and night sweats are common especially in those with extensive disease.

Stages of tuberculous (TB) lymphadenitis

1. Stage of lymphadenitis (Fig. 2.3)

- Common in young adults between 20 and 30 years of age.
- Upper anterior deep cervical nodes are enlarged.
- Nontender, discrete, mobile, firm lymph nodes are palpable.

2. Stage of periadenitis/Stage of matting (Fig. 2.4)

- Results due to involvement of capsule
- Nodes move together
- Firm, nontender
- ***Matting is pathognomonic of tuberculosis.***
- Other rare causes of matting are chronic lymphadenitis and anaplastic variety of lymphoma.

3. Stage of cold abscess (Figs 2.5 to 2.9)

It occurs due to caseating necrosis of lymph nodes resulting in fluctuant swelling in the neck. Clinical features of cold abscess in the neck are:

- No local rise of temperature
- No tenderness
- No redness
- Soft, cystic and fluctuant swelling
- Transillumination is negative
- On sternocleidomastoid contraction test, it becomes less prominent indicating that it is deep to the deep fascia.

PEARLS OF WISDOM

Multiple matted mobile lymph nodes are characteristic of tuberculosis.

Fig. 2.5: TB lymphadenitis—cold abscess

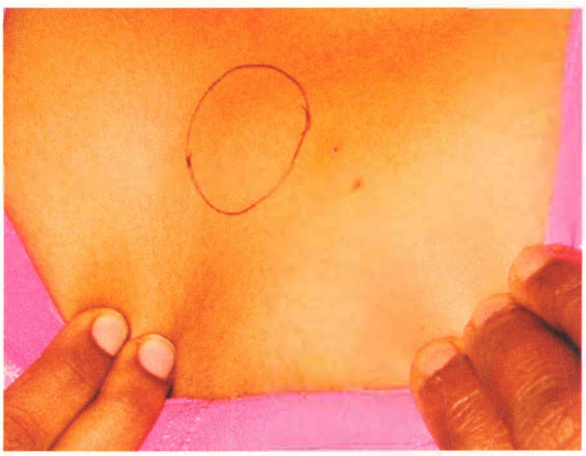


Fig. 2.6: Cold abscess in the second intercostal space

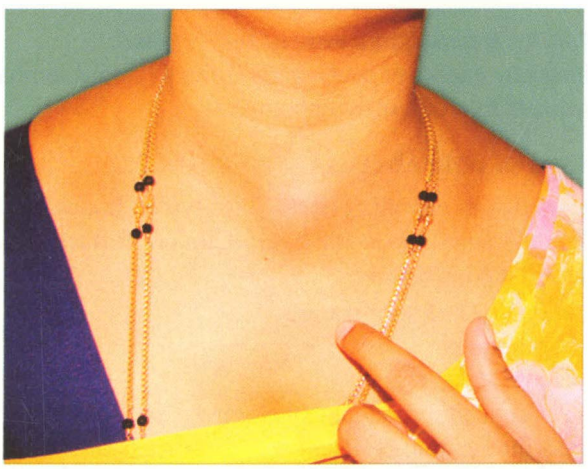


Fig. 2.7: Cold abscess in the midline above suprasternal notch—another common site

Differential diagnosis

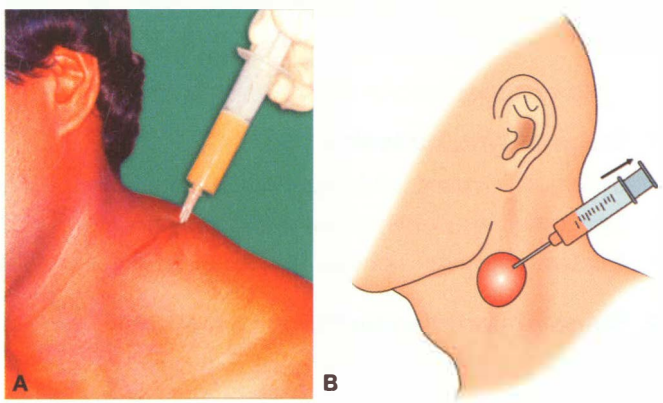
Branchial cyst can be confused for cold abscess in the anterior triangle. Branchial cyst is of longer duration and patients with cold abscess may have other lymph nodes in the neck.

Fig. 2.8: Cold abscess of the right sternoclavicular joint

Fig. 2.9: Cold abscess in the paravertebral region secondary tc tuberculosis of spine

Treatment of cold abscess (Figs 2.10A and B)

- **Nondependent aspiration** by using a wide bore needle to avoid a persistent sinus.
- Wide bore needle is preferred because caseous material is thick.
- Incision and drainage should not be done as it causes persistent tuberculous sinus.
- Antituberculous treatment is given.



Figs 2.10A and B: Nondependent aspiration

4. Stage of collar stud abscess (Figs 2.11 and 2.12)

It results when a cold abscess which is deep to the deep fascia ruptures through the deep fascia and forms another swelling in the subcutaneous plane which is fluctuant. Cross fluctuation test may be positive. It is treated like a cold abscess.

5. Stage of sinus (Fig. 2.13)

- Sinus is a blind tract leading from the surface down into the tissues.
- It occurs when collar stud abscess ruptures through the skin.
- Tubercular sinus is the commonest sinus in the neck in India.

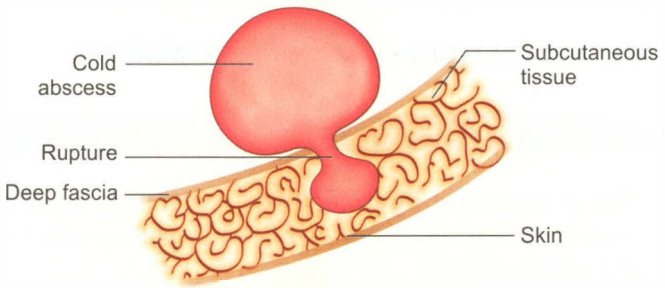


Fig. 2.11: Collar stud abscess

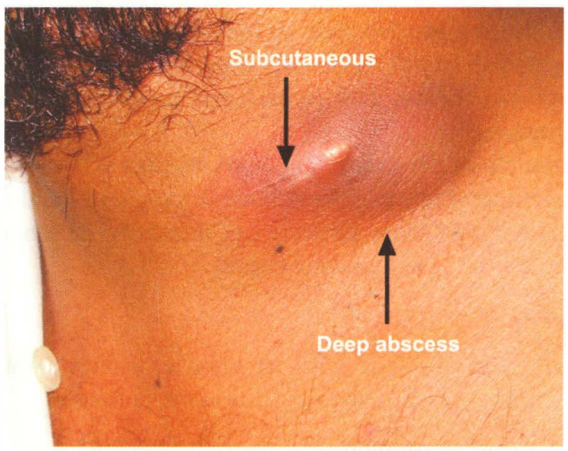


Fig. 2.12: Collar stud abscess

Fig. 2.13: Multiple tubercular sinuses in the neck and in front of the ear

- It is common in young females.
- It can be multiple.
- Tubercular sinus will have a wide opening.
- Resembles an ulcer with undermined edge.
- No induration.
- Skin surrounding the sinus shows pigmentation and sometimes, it is bluish in colour.
- A group of lymph nodes is usually palpable underneath the sinus.

Tuberculosis of Intrathoracic nodes

- Incidence is about 25% in all cases of TB lymphadenitis.
- Pressure on bronchus gives rise to atelectasis, lung infection
- Pressure on oesophagus causes dysphagia, oesophago–tracheal fistula (Fig. 2.14).
- Retroperitoneal nodes may give rise to chylous ascites, chyluria.
- HIV infection and lymph node TB (Key Box 2.4)

Investigations in tubercular lymphadenitis

- Complete blood picture may reveal low Hb%.
- ESR is elevated in majority of cases.
- Chest X-ray is usually negative, also sputum for AFB (acid-fast bacilli).
- FNAC (fine needle aspiration cytology) can give a diagnosis in about 75% of cases.

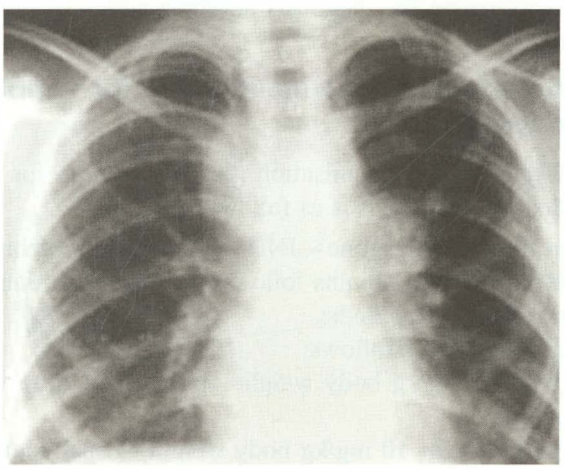


Fig. 2.14: Large mediastinal lymph nodes

KEY BOX 2.4

HIV INFECTION AND LYMPH NODE TUBERCULOSIS

- It is more common than lymphoma and sarcoma
- Patients are male and older
- Multiple sites are more commonly affected
- Disseminated disease (virulent) may be seen
- Nodes can be tender
- Weight loss is more common

Table 2.3 Summary of the management of various stages of tuberculous lymphadenitis

Stage	Investigations	Treatment
Lymphadenitis	Lymph node biopsy/FNAC	Antituberculous treatment (ATT)
Periadenitis	Lymph node biopsy/FNAC	Antituberculous treatment
Cold abscess	Aspirate cheesy material for AFB	Nondependent aspiration with ATT
Collar stud abscess	Aspirate for AFB	Nondependent aspiration followed by ATT
Sinus	Edge biopsy of the sinus	ATT

- Lymph node biopsy reveals central caseation surrounded by epithelioid cells with ***Langhans*** type of giant cells. Langhans type of giant cells usually have more than 20 nuclei. It is gold standard.
- If it is a cold abscess, aspiration will reveal **cheesy material**. AFB is usually negative.
- Edge biopsy from the sinus (may show granuloma).

Pathological types

Caseating type: Most common type seen in young adults.
Hyperplastic type: Lymph nodes show marked degree of lymphoid hyperplasia. Least caseation is seen in patients with good body resistance.
Atrophic type: Seen in elderly patients. Lymphoid tissue undergoes degeneration. Glands are small with early caseation.

Treatment (Table 2.3)

After confirming the diagnosis antituberculous treatment is given.

Antituberculous treatment (ATT) for lymphatic tuberculosis

The World Health Organisation recommendation for extra-pulmonary tuberculosis is as follows:

- The three-drug regime—INH, rifampicin, pyrazinamide (HRZ)—for two months followed by INH and rifampicin for another four months.
- The dosage is as follows:
  - **INH:** 6 mg/kg body weight—usual adult dose is 300 mg/day.
  - **Rifampicin:** 10 mg/kg body weight—usual adult dose is 450–600 mg/day.
  - **Pyrazinamide:** 30 mg/kg body weight—usual adult dose is 1,500 mg/day.
- Detailed dosage of ATT, side effects of these drugs are discussed in medicine textbooks.

Role of surgery in tuberculous lymphadenitis

- **Biopsy:** Lymph node biopsy, wedge biopsy from the edge of the sinus.
- **Aspiration:** Nondependent aspiration of cold abscess and pus should be sent for AFB staining and Ziehl Neelsen (ZN) stain.
- **Excision of lymph nodes** if they persist in spite of antituberculous treatment.
- **Excision of sinus wall** along with the tract.

Other special types of pyogenic infections

BOIL (Key Boxes 2.5 and 2.6)

- This is also called furuncle. It is a hair follicle infection caused by *Staphylococcus aureus* or secondary infection of a sebaceous cyst.
- It starts with a painful indurated swelling with surrounding oedema. After about 1–2 days, softening occurs in the centre and a pustule develops which bursts spontaneously discharging pus. Necrosis of subcutaneous tissues produces a greenish slough. Skin overlying the boil also undergoes necrosis. ***Hence, boil is included under acute infective gangrene.***
- Furuncle of the external auditory meatus is a very painful condition because of the rich nerve supply of the skin. Pain is also due to dense adherence of skin to the perichondrium (there is no subcutaneous tissue).

Treatment of boil

Incision and drainage with excision of slough. Antibiotic cloxacillin is given. Diabetes, if present, is treated.

KEY BOX 2.5

PRECIPITATING FACTORS	COMMON LOCATIONS
• Scratching	Face and back of the neck
• Diabetes	Axilla
• Poor immunity	Gluteal region

KEY BOX 2.6

FACTS ABOUT A BOIL

• Dangerous boil	: On the skin of face
• Tender boil	: External auditory
• Sweet boil	: Diabetic patients
• Boil likes	: Oily skin
• Blind boil or dull boil	: Subsides without suppuration

Complications of boil

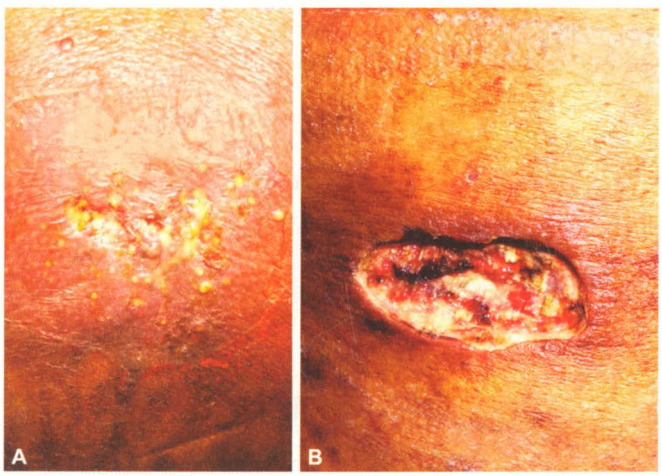
- Necrosis of the skin
- Pyaemic abscess and septicaemia.
- **Cavernous sinus thrombosis** due to boil on the face or stye on the eyelid.

CARBUNCLE (Figs 2.15A and B)

- This is an **infective gangrene** of the subcutaneous tissue caused by ***Staphylococcus aureus*** (Key Box 2.7). It commonly occurs in **diabetic patients**. Patients with poor immunity, or undergoing radiotherapy can also develop carbuncle.
- **Sites: Nape of the neck** is the commonest site followed by back and shoulder region. Skin of these sites is **coarse** and has **poor vascularity.**

Pathology

The initial lesion is similar to a boil in the form of hair follicle infection with perifolliculitis. Since majority of patients are diabetics, infection takes a virulent course and results in necrosis of subcutaneous fat which gives rise to multiple abscesses. These abscesses are **intercommunicating** and they open to the exterior by multiple openings which are called **sieve-like openings.** This appearance is described as **cribriform** appearance which is pathognomonic of carbuncle.



Figs 2.15A and B: (A) Carbuncle of the back of neck—common site, sieve like appearance, (B) after excision of the carbuncle—wound heals within 2 to 3 weeks. Some cases require split skin grafting

KEY BOX 2.7

STAPHYLOCOCCAL INFECTIONS OF SURGICAL IMPORTANCE

- Boil
- Carbuncle
- Breast abscess
- Parotitis
- Osteomyelitis

Clinical features

- Typically, the patient is a diabetic.
- Severe pain and swelling in the nape of the neck.
- Constitutional symptoms such as fever with chills and rigors are severe.
- Surface is red, angry looking like **red hot coal**.
- Surrounding area is indurated.
- Later, skin on the centre of carbuncle softens and peripheral satellite vesicles appear, which rupture discharging pus and giving rise to **a cribriform** appearance (Fig. 2.15A).
- The end result is development of a large crateriform ulcer with central slough.

Complications

1. Worsening of the diabetic status resulting in diabetic ketoacidosis.
2. Extensive necrosis of skin overlying carbuncle. Hence, it is included under acute infective gangrene.
3. Septicaemia, toxaemia.

Treatment (Key Box 2.8)

- Diabetes control, preferably with injectable **insulin**.
- Appropriate **parenteral antibiotics** are given till complete resolution occurs. Most strains of *Staphylococcal aureus* are sensitive to cloxacillin, flucloxacillin, erythromycin and some of the cephalosporins. However, ***methicillin-resistant Staphylococcal aureus (MRSA)*** bacteria are resistant to the drugs mentioned above. They are sensitive only to expensive drug **vancomycin** which has to be given intravenously.
- Improve general health of the patient.
- If carbuncle does not show any softening or if it shows evidence of healing, it is not incised. It can be left open to the exterior or saline dressings may be applied to reduce oedema. Complete resolution may take place within 10–15 days.
- Surgery is required when there is pus. ***Cruciate incision*** is preferred because of multiple abscesses and extensive subcutaneous necrosis. Edges of the skin flap are excised, pus is drained, loculi are broken down, slough is excised, and ***cavity is irrigated with antiseptic agents***. Like pyogenic

KEY BOX 2.8

SUMMARY OF CARBUNCLE

- Caused by **C**occi
- Abscesses **C**ommunicating
- Red hot like **C**oal
- Appearance **C**ribriform, Crateriform ulcer
- Gangrene **C**utaneous (subcutaneous)
- Drug of choice **C**loxacillin
- Diabetes **C**ontrol
- Incision **C**ruciate

Observe **8 Cs**

Fig. 2.16: Central slough and cruciate incision followed by excision should be done

abscess, wound heals with granulation tissue from the depth (Fig. 2.16)—excision of carbuncle.

ERYSIPELAS

- It is an acute inflammation of the skin and subcutaneous tissue associated with severe lymphangitis. Causative organism is *Streptococcus pyogenes*. Precipitating factors are malnourishment, chronic diseases, etc. Thus, children and old people are commonly affected.
- Infection sets in after a small scratch or abrasion and spreads very rapidly resulting in toxaemia. Sites: Face, eyelids, scrotum and in infants, the umbilicus.

Clinical features

- **Rose-pink** rash with raised edge, appreciated on palpation and has a consistency of button hole.
- **Vesicles** appear later, rarely become pustular.
- Oedema of the eyelids or scrotum depending upon the site.
- Features of ***toxaemia***
- When it occurs in the face, it involves pinna because erysipelas is basically a ***cuticular lymphangitis***. This is described as ***Milian's ear sign positive***. This sign is used to differentiate cellulitis of face from facial erysipelas. In cellulitis of face, pinna does not get involved because of close adherence of skin to the cartilage.

Complications

1. Toxaemia and septicaemia
2. Gangrene of skin and subcutaneous tissue
3. Lymphoedema of face and eyelids due to lymphatic obstruction causing fibrosis of lymphatics.

Treatment

- Injection crystalline penicillin 10 lakh units 6th hourly IM/IV for 5–10 days.

CHRONIC ABSCESS

It occurs when the initial infective process or the cause is not fully identified and properly treated.

Sites

Foot, hand, thigh, etc.

Causes

1. **Foreign bodies:** These are the most common causes for chronic abscess. Typical history of a recurrent swelling discharging pus is present. Wooden pieces impacted in the thigh or in the foot are common. Synthetic mesh used in repair of hernias getting infected is another example.
2. **Dead tissue:** As it occurs in diabetic patients.
3. **Pilonidal sinus:** This condition gives rise to recurrent abscesses. Typical history of pain and swelling which ruptures followed by spontaneous recovery is present. However, the sinus persists.
4. **Chronic disease:** **Tuberculosis** is one of the causes. All features of cold abscess can be present but in an unusual location.

CLINICAL NOTES

A 40-year-old female presented with swelling of the left thigh of 8 months duration. There were no signs of inflammation. FNAC was inconclusive. At surgery, thick-walled, localised abscess with fleshy tissue was removed. The final report was tubercular abscess.

There was no evidence of tuberculosis anywhere in the body. In many cases, tuberculosis can present in different forms as in this case. Detailed investigations could not reveal any evidence of pulmonary tuberculosis.

NECROTISING FASCIITIS

It is a spreading, destructive, invasive infection of skin and soft tissues including deep fascia with ***relative sparing of muscle***.

Common sites

It is common in lower extremities. Other sites are genitalia, groin, lower abdomen. In these places it is comparable or similar to gangrene and is called **Meleney's** gangrene. Other sites are the lower extremities.

Causative organisms

Two types have been identified:

- **Monomicrobial:** It is due to group A *β-haemolytic streptococci*. It is also called **type II necrotising fasciitis**.

- **Polymicrobial:** It is due to synergistic combination of *anaerobes* and *coliforms* or *nongroup A streptococci*—**type I necrotising fasciitis**.
- Very often there is **no history of injury** when it occurs in the lower limbs.

Risk factors for type I necrotising fasciitis
(Key Box 2.9)

KEY BOX 2.9

NECROTISING FASCIITIS—RISK FACTORS

- Diabetes mellitus, malnutrition
- Obesity, corticosteroids
- Immune deficiency

Clinical features

- Sudden pain in the affected area with gross swelling of the limbs (Figs 2.17 to 2.19).
- The part is swollen, red, erythematous and oedematous with skip lesions of skin necrosis and ulceration.
- ***Skin changes:*** Bronze hue, brawny induration, blebs or crepitus are other important features.
- High degree fever, jaundice, renal failure can occur soon in untreated cases (Key Box 2.10).

KEY BOX 2.10

SPECIFIC FEATURES OF TYPE II NECROTISING FASCIITIS

- Caused by *Streptococcus pyogenes*
- Occur in young healthy people
- Minor abrasions, laceration may be a precipitating factor
- Severe systemic illness with multiorgan failure—Streptococcal toxic shock syndrome

Diagnosis

Full thickness biopsy taken at bedside can give full diagnosis. Watery pus (**dishwater liquid**) is also a characteristic.

Treatment

Early, aggressive treatment includes supportive and surgical treatment.

Supportive treatment

- This includes hospitalisation, adequate hydration, broad-spectrum antibiotics. **Vancomycin with carbapenem** may be required urgently.
- Surgery involves wide excision, generous debridement followed by skin grafting, a few days or weeks later.
- **In type II cases (streptococcal):** High dose penicillins along with clindamycin is the treatment of choice.

Fig. 2.17: Type II necrotising fasciitis in a healthy man. In spite of debridement and intensive care, he died of multiorgan failure

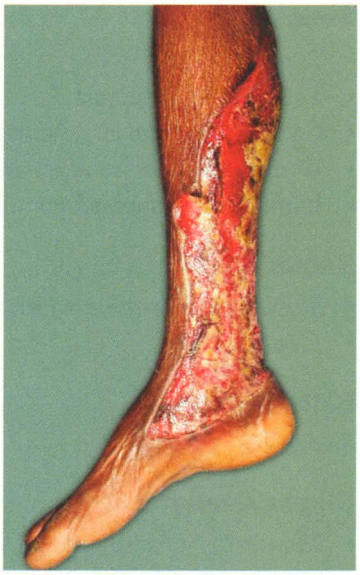


Fig. 2.18: This patient presented with features of cellulitis with renal failure. There were no precipitating factors. After debridement, recovery was complete

Fig. 2.19: Observe the skin changes in necrotising fasciitis

Clindamycin has special effect as it is a **potent suppressor of bacterial toxin synthesis**.

ACUTE PYOMYOSITIS

Definition

Localised area of suppuration **within striated muscle** is pyomyositis.

Aetiology

- Trauma
- Transient bacteraemia
- Common in tropical countries
- IV drug abusers are often affected
- Immunocompromised conditions

Bacteriology

- *Staphylococcus aureus* (90%), *Streptococcus pyogenes*
- *E.coli*

Pathogenesis

See Key Box 2.11

Clinical features

- Classically quadriceps, gluteus, shoulder and upper arm muscle are affected. Pain over the part, oedema, fever, jaundice are common. Tenderness, induration and spasm of muscles are characteristic.
- **Renal failure follows soon.**

Investigations

- Sonographic aspiration of pus followed by culture
- CT, MRI are ideal investigations to know the spread of the infection.
- Creatine kinase can go up to 50,000 to 2,00,000 units/L during acute phase due to **rhabdomyolysis**.

Treatment

- Early diagnosis and early aggressive treatment
- Antibiotics
- Exploration—for diagnosis and as a treatment
- Wide excision of muscles and compartmental excision till viable tissues are visible (Figs 2.20 and 2.21).

Summary—Key Box 2.12.

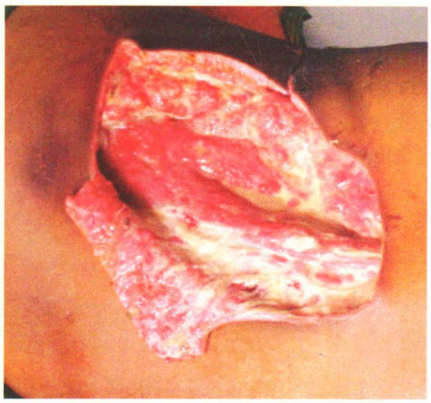


Fig. 2.20: Extensive pyomyositis affecting muscles of the back

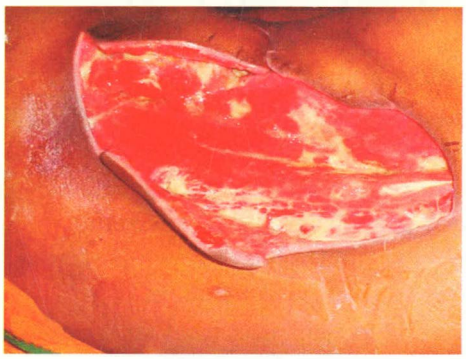


Fig. 2.21: Healing after 2 weeks

KEY BOX 2.11

PATHOGENESIS OF ACUTE PYOMYOSITIS

Trauma
↓
(Striated muscles are rich in iron)
↓
Release of iron
↓
Profuse growth of bacteria
↓
Intramuscular abscess
↓
Spreading infection
↓
Septicaemia

NOSOCOMIAL INFECTIONS

- ***Acquired infection from the hospital is called nosocomial infection.***
- Infection can be from the patient's own organisms (self-infection) or organisms from external sources.
- ***Surgical site infections (SSIs) are the third most frequently reported nosocomial infection,*** first one being pneumonia and the second, catheter-related (including urinary tract infection).

KEY BOX 2.12

PYOMYOSITIS

- **T**rauma
- **T**ransient bacteraemia
- **T**ropical countries
- **T**high muscles are affected
- **T**ender intramuscular abscess
- **T**enderness, temperature, toxicity
- **T**otal renal failure—rhabdomyolysis
- **T**reatment—early aggressive exploration and excision

Observe **8 Ts**

SURGICAL SITE INFECTIONS (SSI)

Common sources of infections

SSIs are divided into incisional superficial (skin and subcutaneous tissue), incisional deep (fascial and muscles) and organ/space related, e.g. intra-abdominal abscess.

In surgical wards, discharging wounds, infected urine, faeces, sputum are all sources of nosocomial infection.

Organisms

Staphylococcus aureus remains the most common pathogen in SSIs followed by coagulase negative staphylococci, enterococci and *Escherichia coli.*

Main sources of SSIs (Key Box 2.13)

1. **Environment:** This is from the operating room. This can be prevented by maintaining ***positive pressure*** in operating rooms, mechanical ventilation with air conditioning providing filtered air through the filter beds, maintaining temperature ***20–22°C***, etc.
2. **Patient-related:** Preoperative bath/shower and medicated soap (or chlorhexidine) is recommended. Preoperative ***hair removal (clipping) should be done immediately before an operation***. Patient's skin is prepared by iodophors (povidone iodine 10%) or chlorhexidine gluconate 4% in alcohol. Povidone iodine is safe, fast-acting, broad-spectrum with some sporicidal activity.
3. Colony of microorganisms increases in intensive care unit patients on previous antibiotic therapy, preoperative shaving, prolonged surgery and remote site infections.

Bacterial factors

• Klebsiella	Surface capsule	Inhibit phagocytosis
• *Streptococcus pneumoniae*	Surface capsule	Inhibit phagocytosis
• Coagulase negative staphylococci	'Slime, on the surface	
• Gram-negative bacteria	Endotoxins or lipopolysaccharide	
• Clostridia, streptococci	Powerful exotoxins	

4. Surgeon and team ***should scrub at least for 3–5 minutes*** with 4% chlrohexidine gluconate. However, chlorhexidine ***alcohol-based 'fast rub'*** is more popular now.
5. **OT attire and drapes:** Scrub suits, caps, masks, double gloves and dedicated footwear are used as barriers. Life of a sterile glove is 3 hours. It should be changed if surgery lasts for more than 3 hours.
6. **Good surgical technique:** Gentle tissue handling, perfect haemostasis, removal of dead tissues, appropriate use of sutures, drains and antibiotics play a major role in preventing SSI.

KEY BOX 2.13

RISK FACTORS FOR DEVELOPMENT OF SURGICAL SITE INFECTIONS

- **Patient-related** (*Remember as* **PATIENT**)
  - **P**eripheral vascular disease and smoking
  - **A**naemia
  - **T**rauma
  - **I**mmunosuppression including diabetes
  - **E**lderly (old age)
  - **N**utritional: Malnutrition
  - **T**oo much obesity
- **Local features** (*Remember as* **LOCAL**)
  - **L**ong (prolonged) surgical procedure
  - **O**xygenation is poor (hypoxia)
  - **C**ontamination of instruments and skin (poor skin preparation)
  - **A**ntibiotic prophylaxis is inadequate
  - **L**ocal tissue necrosis and low temperature **(hypothermia)**

PEARLS OF WISDOM

Blood sugar level < 150 mg/dl in the perioperative period is recommended to decrease morbidity and mortality.

Prevention of hospital infection

- Avoid unnecessary antibiotics to prevent development of resistant organisms (Fig. 2.22)
- Autoclaving and sterilisation should be optimally done
- Proper ventilation of the wards
- Proper scrubbing before any procedure
- Proper disposal of urine, faeces, sputum
- Use of disinfectants.
- Antibiotic prophylaxis (Key Boxes 2.14 and 2.15).

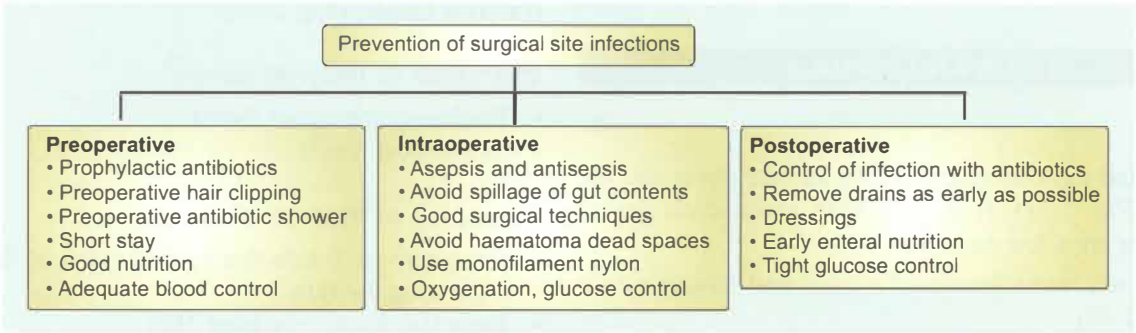


Fig. 2.22: Prevention of surgical site infections

KEY BOX 2.14

ANTIMICROBIAL PROPHYLAXIS

Operation	Likely pathogens
• Breast	*S. aureus*, coagulase-negative Staphylococci
• Appendicectomy	Gram-negative bacilli, anaerobes
• Biliary tract	Gram-negative bacilli, anaerobes
• Upper GI	Gram-negative bacilli, Streptococci, oropharyngeal anaerobes (pepto-streptococci)

Cefazolin is generally accepted as the antimicrobial agent of choice for clean-contaminated operations.

- Dose: 1–2 g/adult dose
- Timing: No more than 30 minutes before skin is incised.

PROPHYLACTIC REGIMENS

- **Vascular:** 3 doses of flucloxacillin with or without gentamicin, vancomycin
- **Oesophagogastric:** 1–3 doses of 2nd generation cephalosporin
- **Biliary:** One dose of 2nd generation of cephalosporin
- **Small bowel:** 1–3 doses of 2nd generation of cephalosporin with metronidazole

KEY BOX 2.15

KEY POINTS IN SSI

- SSI within 24 hours is caused by clostridia and streptocci
- SSI after 48 hours (5 days) is caused by gram –ve and other bacteria.
- Prevention of SSI is by aseptic and antiseptic technique in OT as introduced by Lister, use of prophylactic antibiotics and the third is patient's own ability to prevent infection.
- Skin to be prepared by germicidal antibiotics such as tincture of iodine, povidone iodine or chlorhexidine.
- The first dose of prophylactic antibiotics are given intravenously at the induction of anasthesia.
- Monofilament sutures are better to decrease the SSI.

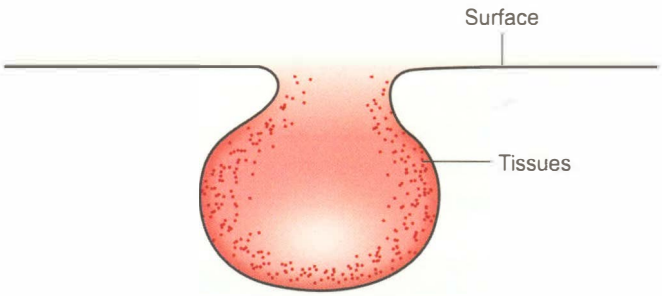


Fig. 2.23: Sinus (blind track)

ASEPSIS AND ANTISEPSIS

Strictly speaking, they are equivalent and there is not much of a difference between these.

Asepsis means precautions taken before any surgical procedure against development of infection. Some examples are: *Wearing gloves before any procedure, cleaning the patient's abdomen with iodine and spirit, sterilisation of instruments and* autoclaving.

Antisepsis: All surgical procedures today are done after taking aseptic precautions.

- Dressing of an already contaminated wound using carbolic acid, iodine.
- Broad-spectrum antibiotics are used in presence of infection.
- Wearing mask and cap in the operation theatre.

SINUS AND FISTULA

SINUS

- It is a blind track leading from the surface down into the tissues (Fig. 2.23). It is lined with granulation tissue. Following are a few examples:
  1. *Congenital sinus*: Preauricular sinus, post-auricular sinus (Fig. 2.25).
  2. *Acquired sinus*:
     - *Median mental sinus* (*see* page 295): Occurs as a result of tooth abscess.
     - *Pilonidal sinus*: Occurs in the midline in the anal region (Fig. 2.25).
     - *Osteomyelitis*: Gives rise to sinus discharging pus with or without bony spicules (Figs 2.26 and 2.27)
- Most common sinus in the neck is due to tubercular lymphadenitis. It discharges cheesy material. Skin surrounding the sinus shows bluish discolouration.

FISTULA

It is an abnormal communication between the lumen of one viscus and the lumen of another (internal) or communication of one hollow viscus with the exterior, i.e. body surface (external fistula) (Fig. 2.24).

Examples of internal fistula

- Tracheo-oesophageal fistula
- Colovesical fistula

Examples of external fistula

- Orocutaneous fistula due to carcinoma of the oral cavity infiltrating the skin
- Branchial fistula (*see* page 253)
- Thyroglossal fistula (*see* page 248)

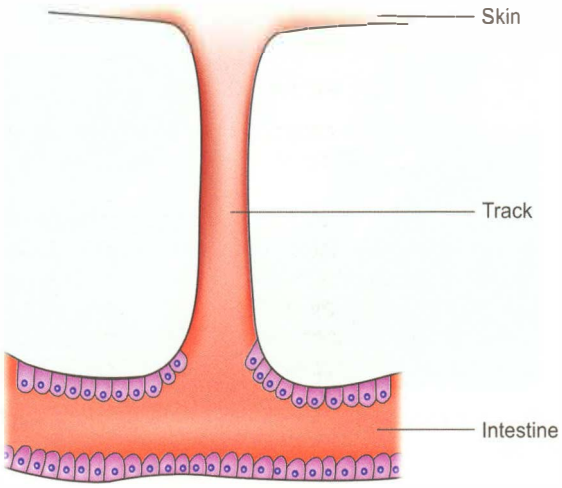


Fig. 2.24: Enterocutaneous fistula (communication)

Causes of persistence of a sinus or fistula

1. Presence of foreign body
2. Persistent infection
3. Distal obstruction as in enterocutaneous fistula
4. Absence of rest
5. Epithelialisation of the track
6. Malignancy
7. Nondependent drainage, inadequate drainage
8. Dense fibrosis
9. Irradiation
10. Specific causes—tuberculosis, actinomycosis.

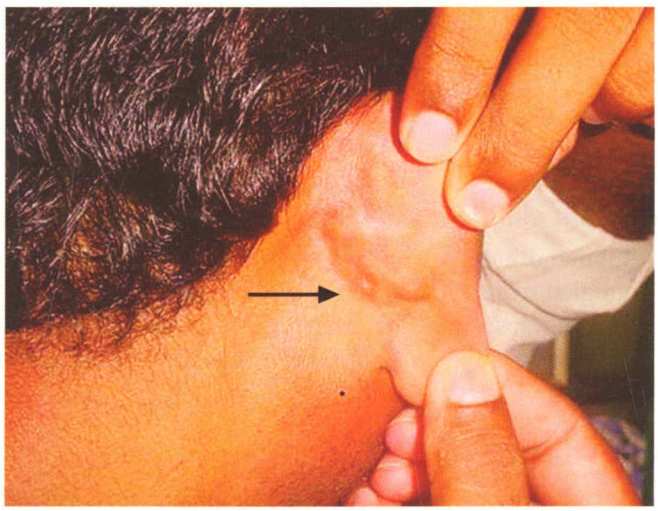


Fig. 2.25: Postauricular sinus

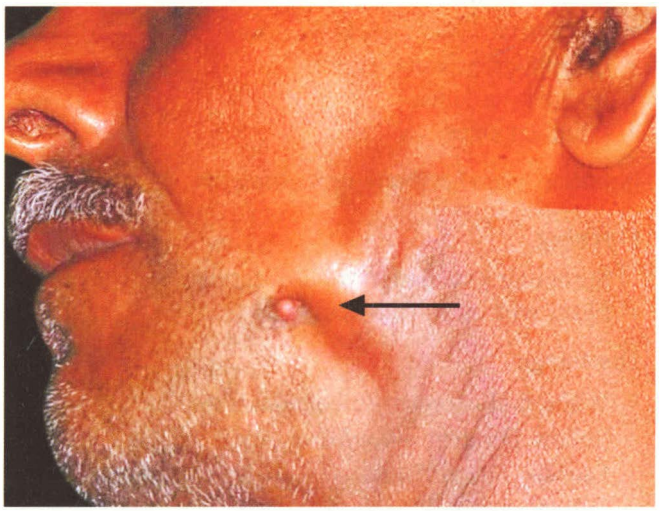


Fig. 2.26: Mandibular sinus due to badly infected caries teeth, osteomyelitis of mandible

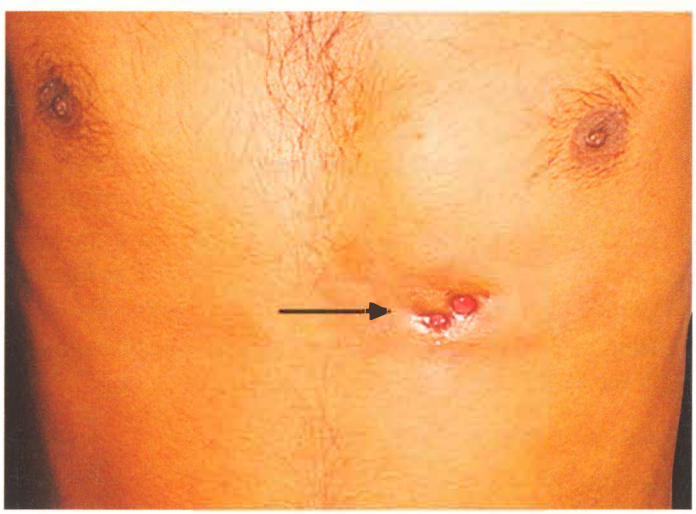


Fig. 2.27: Tuberculous sinuses in the chest wall. Observe that the edge of the sinuses are in flush with the skin

Fig. 2.28: A patient with pilonidal sinus positioned in jack-knife position for excision

CLINICAL EXAMINATION OF SINUS AND FISTULA

Inspection

- **Location** gives the diagnosis in the majority of the cases of sinus or fistula.

  Fistulas

  - Branchial fistula: Anterior border of lower third of sternomastoid
  - Parotid fistula: In the parotid region
  - Thyroglossal fistula: Midline of neck below hyoid bone
  - Appendicular fistula: Right iliac fossa

  Sinuses

  - **Preauricular sinus:** Front of root of helix of ear due to failure of fusion of ear tubercles. Direction of the sinus is upwards and backwards
  - Median mental sinus—symphysis menti
  - Tubercular sinus—neck/chest wall (Fig. 2.27)
  - Lymphogranuloma—groin
  - Mandibular sinus
  - Pilonidal sinus

- **Number**—can be single or multiple

- **Opening**
  - Sprouting granulation tissue—foreign body
  - Flush with skin—tuberculosis

- **Discharge**
  - White thin caseous—tuberculosis
  - Yellow purulent—*staphylococci*
  - Faecal—faecal fistula
  - Yellow granules—actinomycosis
  - Thin mucus discharge—branchial fistula
  - Urine—urinary fistula.

- **Surrounding skin**
  - Red, angry looking—inflammatory
  - Bluish discolouration—tuberculosis
  - Pigmentation—chronic sinus
  - Skin excoriation—faecal fistula

Palpation

- Temperature and tenderness is increased if there is inflammation of the sinus, e.g. pilonidal sinus.
- Discharge after application of pressure. It suggests nature of fluid.
- **Induration is present in chronic fistula,** actinomycosis, osteomyelitis, etc.
- In tubercular sinus, induration is absent.
- **Fixity:** Osteomyelitis sinus is fixed to the bone and median mental sinus may be fixed to the jaw bone. Test is done by trying to move the sinus.

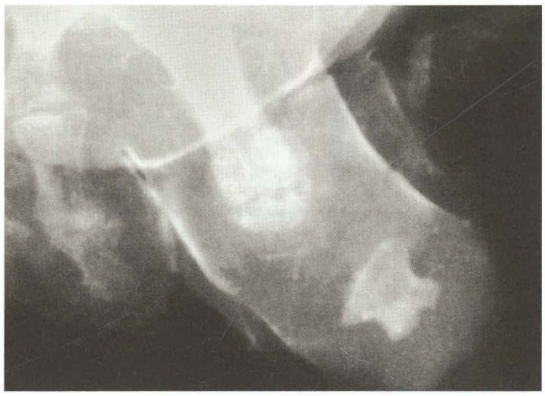


Fig. 2.29: Persisting sinus due to osteomyelitis of distal phalanx of great toe

- **Probing** to know the depth of sinus is not recommended.
- **Palpation at a deeper plane**
  - Enlarged nodes in tuberculosis or lymphogranuloma venereum
  - Thickening of mandible or bone
  - Submandibular stone may be palpable as in submandibular fistula.

Relevant clinical examination

- Submandibular gland enlargement can be detected by bidigital examination.
- Alveolar abscess can be found as in median mental sinus.
- Per rectal examination and proctoscopy may reveal internal opening of the fistula.

Investigations

- Complete blood picture (**CBP**—Haemoglobin %, total and differential count, erythrocyte sedimentation rate—**ESR**): **ESR** may be increased as in tuberculosis. Increased total count suggests infection.
- Urine sugar, fasting blood sugar **(FBS)** and postprandial blood sugar **(PPBS)** to rule out diabetes.
- **X-ray of the part:** To look for osteomyelitis of mandible, toe and also, for any foreign body (Fig. 2.29).
- **X-ray kidney, ureter, bladder region (KUB), ultrasound abdomen:** Staghorn calculi as in lumbar urinary fistula.
- **Fistulography or sinusography** is done to know the exact extent or origin of the sinus or fistula. Dye such as **lipoidal (poppy seed oil contains 40% iodine)** is used.
- **Biopsy from the edge of sinus** is done if specific aetiology is suspected, e.g. tuberculosis, malignancy.

Management (Key Box 2.16)

Following are a few examples:

- Sequestrectomy for osteomyelitis
- Control of tuberculosis for tubercular sinus in the neck.
- Removal of the foreign body if present (clinical notes)

KEY BOX 2.16

BASIC PRINCIPLES

- Antibiotics
- Adequate rest
- Adequate excision
- Adequate drainage

- If the track is well formed and epithelialised, entire track has to be removed even if the disease is under control.

Please note: Details regarding individual fistula and sinus have been discussed in their respective chapters.

MISCELLANEOUS

IRIS: Tuberculosis associated **I**mmune **R**econstitution **I**nflammatory **S**yndrome. This is seen in patients with tubercular lymphadenitis on retroviral therapy. It manifests as deterioration of a treated infection or new presentation of previously subclinical infection. Thus lymph node may persist or may become bigger.

- It occurs due to antigenic response due to bactericidal action of anti-tubercular drugs.
- Needs only reassurance, anti-inflammatory drugs.
- Short course of steroids may be needed if pressure symptoms are present.
- However, if lymph node does not respond and if it is increasing, biopsy should be done to rule out lymphoma.

CLINICAL NOTES

1. *A patient who underwent surgery for varicose veins, had persistent seropurulent discharge from the inguinal incision. Initially, it was thought to be due to infection. The discharge persisted for a period of two months. The wound was explored. A gauze piece was found and removed. The wound healed well. Retrospective analysis of the surgery revealed slipping of the ligature applied to the long saphenous vein and several gauze pieces were used to control the bleeding point.*
2. *We had a sixty-year-old man who had a small sinus in the loin discharging watery fluid. He had seen many doctors for many years. He was treated with antibiotics and even antituberculous treatment without any relief. X-ray KUB revealed a staghorn calculus.*

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- More and more importance has been given to surgical site infection (SSI). Strict guidelines must be followed in the operation theatre to decrease the incidence of surgical site infection.
- Surgical site infections are also nosocomial infections.
- **IRIS** has been added

MULTIPLE CHOICE QUESTIONS

1. Treatment of cold abscess is:
A. Excision
B. Incision and drainage
C. Marsupialisation
D. Nondependent aspiration

2. Tubercular sinuses in the neck—following are true *except*:
A. Usually multiple
B. Edge is bluish in colour
C. Induration is very characteristic
D. Jugulo-digastric nodes are commonly affected

3. The ideal treatment of carbuncle is:
A. Drainage
B. Incision and drainage
C. Excision
D. Aspiration

4. Following facts are true in Ludwig's angina *except*:
A. It is caused by *Staphylococcus aureus*
B. Diffuse swelling in the submental and submandibular region is common
C. Putrid halitosis is commonly found
D. It can give rise to mediastinitis also

5. Following are true in pyaemic abscess *except*:
A. Usually they are multiple
B. They are deep to deep fascia
C. Diagnostic feature is high local rise in temperature
D. It occurs due to pyaemic process

6. Following are true for carbuncle *except*:
A. Nape of the neck is the commonest site
B. Cribriform appearance is diagnostic
C. Abscesses are not communicating with each other
D. *Staphylococcus* is the commonest organism

7. Following are true for erysipelas *except*:
A. Rose pink rash is common
B. Cuticular lymphangitis is an important component
C. Pinna never gets affected in facial erysipelas
D. It is caused by *Streptococcus pyogenes*

8. Muscles are spared in which condition?
A. Gas gangrene
B. Necrotising fasciitis
C. Pyomyositis
D. Acute embolic gangrene

9. Following are features of necrotising fasciitis type II *except*:
A. It is monomicrobial—β-haemolytic streptococci
B. It can occur in young healthy individuals also
C. Organism is not sensitive to clindamycin
D. It can give rise to toxic shock syndrome

10. Which of the following is true to prevent surgical site infection?
A. Preoperative hair clipping should be done just prior to surgical procedure
B. Skin is prepared by 20% povidone iodine
C. Blood sugar level should be maintained within 200 mg/dl
D. Positive pressure ventilation at temperature of 25 °C is ideal

ANSWERS

1 D 2 C 3 C 4 A 5 C 6 C 7 C 8 B 9 C 10 A

3

Tetanus and Gas Gangrene

- Tetanus
- Gas gangrene
- Types of gas infections
- A case report of neck rigidity
- What is new?/Recent advances

TETANUS

Introduction

A nonimmunised, eighteen-year-old girl was admitted with moderate tetanus following a nail prick in her foot. In the hospital, she developed convulsions, laryngeal oedema and cardiac arrest from which she was resuscitated and shifted to intensive care unit under **anaesthesiologist's care**. Tracheostomy, ventilation and paralysing agents were used. Unlike many others, she was lucky. She walked home after two months of stay in intensive care unit after a lot of suffering and spending a large amount of money. The case history has been written here to impress upon the students, the following:

1. How important is immunisation to prevent tetanus?
2. How serious is this disorder?
3. Is it possible to save these patients who are critically ill?

Aetiopathogenesis

Tetanus is a serious disorder with very high morbidity and mortality even with treatment. The disease is caused by *Clostridium tetani*, an anaerobic spore-forming bacillus with terminal spore which has a drumstick-like appearance.

PEARLS OF WISDOM

Narcotic addicts who inject themselves beneath the skin at many sites are vulnerable—'Skin Poppers'.

Possible routes of infection

- **Umbilical cord, in neonates**, seen in communities which practise cowdung application on the umbilical stump.
- Wound, as a complication of road traffic accidents where other aerobic organisms reduce oxygen tension in the wound, thereby facilitating growth of anaerobic *Clostridium tetani* (Key Box 3.1).
- **Minor injuries** with **rusted nails**, piercing of the ear lobes, tattooing, injections, etc.
- Endogenous infection after **septic abortion** or **surgical operations** on gastrointestinal tract.
- Tetanus due to infection acquired in the operation theatre.

Thus, tetanus is a wound infection. **"No wound, no**

KEY BOX 3.1

WOUNDS WHICH ARE MORE PRONE FOR TETANUS

Time—wound more than 6 hours old
Extensive contamination by soil, faeces, rust
Tissue devitalised or denervated
Animal or human bites
No less than 1 cm (more than 1 cm)
Ulcer or wound—deeper
Stellate wounds—burst type

Remember as **TETANUS**

tetanus" is true. Having entered the wound, the organisms multiply and produce powerful exotoxins which produce the disease. Thus, the organisms by themselves, do not produce the disease. The toxins produced by the organisms are tetanospasmin (neurotoxin) and tetanolysin (haemolysin).

- **Tetanospasmin has affinity towards nervous tissues.** It reaches the central nervous system along the axons of motor nerve trunks. The toxin gets fixed to motor cells of the anterior horn cells. The toxin, which is fixed to the motor end plate, acts in the following ways:
  1. It **inhibits the release of cholinesterase** leading to accumulation of acetylcholine at the motor end plate. This causes tonic rigidity of the limb, trunk, abdominal and neck muscles.
  2. It acts at the ***spinal level*** and causes reflex contraction of muscles due to minor stimuli.
- **The toxin which is fixed to the nervous tissue cannot be neutralised**. However, the circulating toxin can be neutralised. Incubation period may vary from a few days to months or years. Hence, it is not important. The interval between first symptom (dysphagia and stiffness of jaw) to a reflex spasm is called the **period of onset**. If this is less than 48 hours, the prognosis is poor and if more than 48 hours, prognosis is better.

Favourable conditions for development of tetanus

- No immunisation
- Foreign body
- Injury
- Improper sterilisation
- Devitalised tissues
- Anaerobic conditions.

Special types of tetanus

1. **Tetanus neonatorum:** It occurs due to contamination of umbilical cord in children born to nonimmunised mothers. It manifests usually around 6–8 days of birth and is called **eighth day disease**. It carries almost 100% mortality.
2. **Local tetanus:** In this, contraction of muscles occurs in the neighbourhood of the wound.
3. **Cephalic tetanus:** Usually occurs after wound of head and face. Cranial nerves such as facial nerve and oculomotor nerve can get **paralysed**. It carries poor prognosis.
4. **Bulbar tetanus:** It is a condition wherein muscles of deglutition and respiration are involved. It is fatal.
5. **Latent tetanus:** It develops after a few months to years following a wound which might have been forgotten.
6. **Puerperal tetanus:** It occurs as a complication of abortion or puerperal sepsis.
7. **Postoperative tetanus:** Occurs due to improper sterilisation of instruments and carries 100% mortality. This type of tetanus should not occur, in a modern operation theatre.
8. **Otitis tetanus:** It is due to chronic suppurative otitis media. In these cases, the wound is a tear in the tympanic membrane. It can occur in any age group, but commonly occurs in children and young adults.

Clinical features (Table 3.1)

- **Autonomic dysfunction:** Increased basal sympathetic tone manifesting as tachycardia, bladder, bowel dysfunction, labile hypertension, pyrexia, pallor, sweating and cyanosis of the digits can occur.
- Episodes of bradycardia, low central venous pressure and even cardiac arrests have been reported due to parasympathetic dysfunction.
- They can develop complications such as pneumonia, urinary tract infection, etc.

Treatment of established tetanus

I. General management
II. Specific management.

I. General management

- **Admission and isolation**[1] in a quiet room, to avoid even minor stimuli that may precipitate spasm (Fig. 3.1).
- **Wound care** which includes drainage of pus, excision of necrotic tissue, removal of foreign body and proper dressing. Exudate or pus can demonstrate gram-positive rods.
- **Inj. tetanus toxoid** 0.5 ml to be given IM.
- **Antitetanus serum** (ATS) 50,000 units intramuscular (IM) and 50,000 units intravenous (IV). This should be given only after giving a test dose which consists of diluting a small dose of serum with ten times saline and injecting a small amount in the subcutaneous tissue. It has become less popular due to availability of human antitetanus globulin.

[1]Isolation for tetanus has been misunderstood by surgeons. It is a fact that in majority of the hospitals, tetanus patients are isolated in a remote corner of the hospital, well away from the reach of a skilled person. Many cases die due to convulsions and laryngeal spasm before they are intubated and resuscitated. The critically ill patients are admitted in an intensive care unit under the supervision of anaesthesiologist's care in our institution (Fig. 3.1). Tetanus is not communicable from person to person.

Table 3.1 Clinical features with differential diagnosis

Symptoms and signs	Differential diagnosis
• **Trismus** or **lock jaw,** occurs due to severe contraction of the masseter muscle, resulting in inability to open the mouth. It is the most common symptom of tetanus.	• Alveolar abscess or temporomandibular joint involvement
• **Dysphagia** occurs due to spasm of pharyngeal muscles.	• Tonsillitis
• **Neck rigidity**	• Meningitis
• **Rigidity of back muscles**	• Orthopaedic disorder
• **Risus sardonicus** due to spasm of facial and jaw muscles.	• Anxiety neurosis
• **Generalised convulsions** wherein every muscle is thrown into contraction, with severe clenching of teeth, arched back and extended limbs is described as opisthotonos (bow-like body; hence the name **dhanurvatha**).	• Epilepsy
• Mild temperature and tachycardia	• Sympathetic hyperactivity

Fig. 3.1: Tetanus patient recovering in an intensive care unit

- **Instead of ATS, human antitetanus globulin** is better and safe. It does not cause anaphylaxis. It is given in the dose of 3000 to 4000 units **IV**. No test dose is required.
- **Inj. Crystalline penicillin 10 lakh units every 6 hours** is the drug of choice against *Clostridium tetani*. It might have to be given for a period of 7–10 days.
- **Metronidazole** 500 mg IV 8th hourly for 10 days. It has been shown to be more effective than penicillin.
- After recovery, full immunisation with tetanus toxoid is a must.

II. Specific management

A. Mild cases

- There is only tonic rigidity without spasm or dysphagia. These patients are managed by heavy sedation using a combination of drugs so as to avoid spasm or convulsions. An example of the method of treatment followed in our hospital is given in Table 3.2.
- **Benzodiazepines** and **morphine** act centrally to minimise the effects of tetanospasmin.
- **Chlorpromazine** being an α-receptor blocker, can decrease sympathetic activity. Other α-blockers such as phenoxybenzamine, phentolamine have also been used.
- These drugs are repeated in such a way that the patient receives some sedative every two hours. The dosage of the drugs is adjusted once in 2 or 3 days so as to get the maximum effect of sedation or muscle relaxation.
- Injection diazepam 10 mg, tracheostomy set, resuscitation set which includes laryngoscope and endotracheal tubes should be kept ready by the side of the patient.

B. Seriously ill cases

- They have dysphagia and reflex spasms.
- A nasogastric tube is introduced for feeding purposes and to administer the drugs.
- Tracheostomy, if breathing difficulty arises.

Table 3.2 Method of treatment

Drug	Dosage	Time
Chlorpromazine	50–100 mg	8 am, 2 pm, 8 pm and 2 am
Phenobarbitone	30–60 mg	10 am, 4 pm, 10 pm and 4 am
Diazepam	10–20 mg	12 noon, 6 pm, 12 midnight and 6 am

C. Dangerously ill cases

This group includes patients with major cyanotic convulsions. In addition to continuing sedatives, these patients are paralysed with muscle relaxants (neuromuscular blocking agents) and mechanically ventilated till they recover. One cannot predict the duration of the need for ventilatory support. During this period, supportive therapy such as adequate nutrition, care of the urinary bladder and bowel, frequent change of position to avoid bedsore, have to be given.

Prophylaxis

1. *Tetanus neonatorum* can be prevented by immunisation of the mother with two tetanus toxoid injections, **half ml** IM given in the second trimester of pregnancy.
2. Infants and children are immunised with tetanus toxoid, diphtheria and pertussis vaccine (DPT) three doses at 6, 10, 14 weeks of age. This is called triple antigen. A booster dose is given at 18 months and school going time (5 years), and once in five years 0.5 ml of tetanus toxoid is given to achieve active immunity.
3. Immunised individual who receives a provocative injury is administered a booster dose if he has not been given it in the previous 5 years.
4. Tetanus can be **prevented by giving tetanus antitoxin** in the following situations:
   - Wounds of head, face and penetrating wounds
   - Wounds with contused and devitalised tissues
   - War wounds and road traffic accidents

   In such patients, a dose of 250 units of human antitetanus globulin will give adequate protection.

Causes of death

1. **Aspiration** of pharyngeal contents into the lungs resulting in aspiration pneumonia.
2. **Laryngeal spasm and respiratory arrest** resulting in cardiac arrest.
3. **Autonomic disturbances** resulting in cardiac arrhythmias
4. In some patients, **pacemaker insertion** may help if there is refractory bradycardia.

GAS GANGRENE

It is a highly fatal, rapidly spreading infection caused by clostridial organisms which results in myonecrosis.

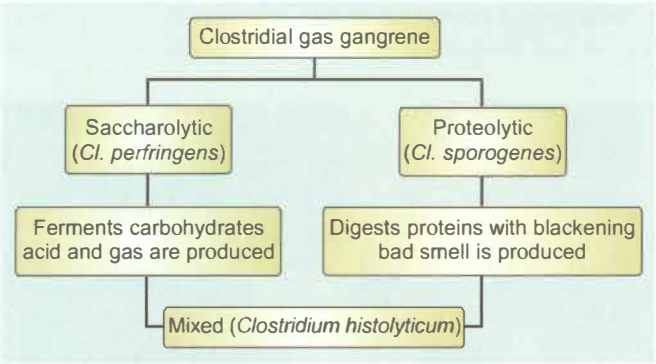
Other names for gas gangrene

Clostridial myositis, clostridial myonecrosis, infective gangrene of the muscles.

Aetiology

The disease is caused by *Clostridium perfringens* (*Clostridium welchii*)—the commonest organism (60%). Other organisms are *Clostridium septicum, Clostridium oedematiens, Clostridium histolyticum.*

These are gram-positive, anaerobic spore-bearing bacilli.



Source of infection (Table 3.3)

Manured soil or cultivated soil, normal intestines.

Risk group

- In patients who have had lower limb amputations performed for ischaemic gangrene, infection can occur from patient's own bowel organisms.
- High velocity gun shot wounds with perforation of hollow viscus are also associated with risk of developing gas gangrene (military wound).
- Immunocompromised patients are at risk.

Pathogenesis (Fig. 3.2)

- Gas gangrene develops in wounds where there is heavy contamination with soil or foreign body, or which is associated with laceration and devitalised muscle mass.

Table 3.3 Predisposing factors for the development of gas gangrene

Factors	Mechanism
Foreign body such as soil, clothing, bullets, glass pieces.	Soil supplies calcium and silicic acid which causes tissue necrosis.
Anoxia due to crushing of the arteries.	Necrosis of the tissues results in proliferation of the organism.
Dead and devitalised tissues.	Anaerobic organisms multiply
Blood clots.	Supplies calcium
Extravasated haemoglobin and myoglobin	Cease to carry oxygen.

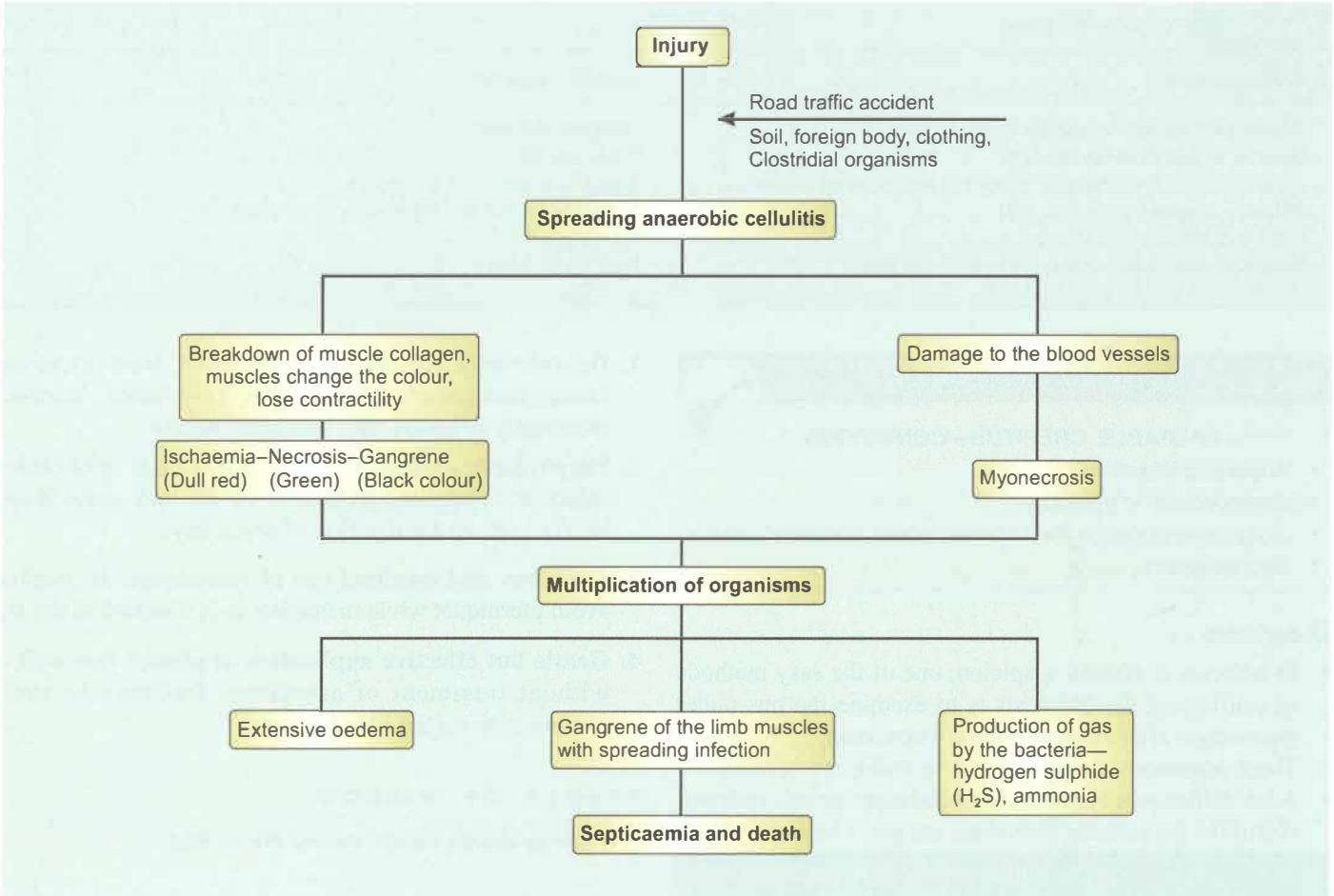


Fig. 3.2: Pathological changes

This type of situation is common today following road traffic accidents. Endogenous infection from patient's faecal matter may be responsible for gas gangrene, in certain cases of contamination of a surgical wound such as below knee amputation done for some other cause. Having entered the wound, *clostridia* multiply and produce powerful toxins.

- All these factors contribute to create a low oxygen tension. Under these favourable conditions clostridial organisms multiply and produce toxins which cause further tissue damage. The toxins produced by the organisms and their effects are given in Table 3.4.
- Once powerful toxins start acting, various pathological events such as inflammation, oedema, muscle necrosis and gangrene of the muscles set in. These events are summarised below.

Clinical features (Table 3.5)

In untreated cases, necrotic process continues and septicaemia, renal failure, peripheral circulatory failure and death occur. Foamy liver is the condition wherein gas is produced in the liver, as a part of septicaemia.

Table 3.4 Toxins and their effects

Toxins	Effects
1. Lecithinase (alphatoxin)—Commonly found in *C. perfringens* type A strain	• Dermonecrosis Haemolysis Profound toxaemia
2. Beta toxin	• Necrosis of the tissues
3. Proteinase	• Breakdown of collagen fibres
4. Hyaluronidase	• Breaks the cement substance of the muscle cells—hyaluronic acid

Table 3.5 Clinical features

Local features	General features
Severe pain and gross oedema of the wound	Anxious and alert
Sutured wound is under tension	Toxic and ill
Thin brownish fluid escapes which has **sickly sweet odour**	Rapid increase in the pulse rate
Palpable **crepitus** (Key Box 3.2)	Hypotension due to suppression of adrenals
Colour changes in the muscles	Vomiting
Skin becomes khaki-coloured due to haemolysis	Low grade fever

KEY BOX 3.2

PALPABLE CREPITUS—CONDITIONS

- Anaerobic infections
- Streptococcal infections
- Surgical emphysema due to oesophageal, tracheal rupture
- Gas gangrene

Diagnosis

- In addition to clinical suspicion, one of the easy methods of confirming the diagnosis is to examine the pus under microscope after staining with Giemsa stain.
- These organisms are gram-positive and spore-bearing.
- A few differences between clostridial myonecrosis and non-clostridial necrotising infections are given below

	Clostridial myonecrosis	Nonclostridial necrotising infections
• Exudate	Thin	Dish water initially purulent later
• Erythema	Absent	Present—mild
• Muscle gangrene	Present	Absent
• Toxicity	Very rapid	Rapid
• Bullae	Haemorrhagic	Nonhaemorrhagic
• White blood cells in the discharge	Absent	Present

- Presence of gas indicates anaerobic metabolism.
- Anaerobic streptococci also produce gas.
- *See* Key Box 3.3

Prophylaxis

Being highly fatal, gas gangrene is better prevented by observing following principles while managing the wound:

KEY BOX 3.3

WHY GAS GANGRENE SPREADS FAST

- Massive infection
- Gross injury
- Devitalised tissue
- Poor immunity
- Foreign body

1. **Debridement:** All dead, necrotic tissue, bone pieces and foreign material are removed. Pus is evacuated. Wound is thoroughly irrigated with antiseptic agents.
2. **Prophylactic antibiotics:** Penicillin is the drug of choice. Injection crystalline penicillin 10–20 lakh units, 4–6th hourly is given for a period of seven days.
3. **Judicious and minimal use of tourniquet:** If possible, avoid tourniquet while managing such a wound in the leg.
4. **Gentle but effective application of plaster cast** with or without treatment of associated fractures to avoid compression on the blood vessel.

PEARLS OF WISDOM

When in doubt, do not suture the wound.

Treatment of established gas gangrene

- **Emergency surgery** which includes excision of all dead muscles and necrotic tissues by using generous, long incisions—debridement.
- **Penicillin** to be continued.
- **Blood transfusions** before, during and after surgery.
- **Polyvalent anti-gas gangrene serum.**
- **Hyperbaric oxygen** will reduce the amount of toxin produced by the organisms (controversial).
- **Do not hesitate to amputate** if it saves the life, because this is the only measure in late cases.
- For summary *see* Key Box 3.4

KEY BOX 3.4

SUMMARY OF GAS GANGRENE

- Correct hypotension
- Control infection
- Treat dehydration
- Early debridement
- Administer hyperbaric oxygen
- Give blood transfusion
- Passive immunisation
- To save life, amputate.

Fig. 3.3: Gas gangrene

CLINICAL NOTES

A 40-year-old gentleman presented to the hospital with massive gas gangrene involving upper limb, chest wall, abdominal wall and back (Fig. 3.3). *It started after an injury to the elbow. The patient received initial treatment in a local hospital. Due to lack of proper facilities, he was neither subjected to any surgical procedure nor was he given any resuscitation. When he came to the hospital, he was in septic shock.*

It was too late when we saw the patient. He was having severe hypotension and renal failure. Resuscitation and debridement was done. However, within six hours of admission to the hospital, he expired.

TYPES OF GAS INFECTIONS

1. **Clostridial cellulitis:** In this condition, healthy muscle is not involved. It involves necrotic tissue and produces features of cellulitis such as tense, swollen parts with palpable crepitus. However, it is a mild infection, which can be managed by antibiotics without surgery (Key Box 3.5).
2. **Local type:** It refers to infection confined to a single muscle.
3. **Group type:** It refers to infection confined to one group of muscles in the compartment. Such cases benefit from a compartmental excision.
4. **Massive type:** Gas gangrene involving the entire limb, needs to be treated by amputation.

KEY BOX 3.5

INFECTIONS BY *CLOSTRIDIUM WELCHII*

- Gas gangrene of the limb
- Gas gangrene of the abdominal wall
- Gangrenous appendicitis, cholangitis
- Necrotising enteritis, food poisoning
- Infection of the uterus following septic abortion.

MISCELLANEOUS

MacLennan anaerobic wound infection

1. Simple wound contamination—no invasion of underlying tissue
2. Anaerobic cellulitis—invasive fascial planes, in muscles, minimal toxins
3. Anaerobic myositis—muscle necrosis.

A case report of neck rigidity

CLINICAL NOTES

A 48-year-old lady underwent vaginal hysterectomy 10 days back and was brought to our hospital, as a case of 'tetanus'. On examination, she had neck rigidity and difficulty in opening mouth. Abdominal rigidity was mild. She was looking pale. Pallor was attributed to anaemia caused by 'dysfunctional uterine bleeding' (DUB), for which she was operated. Diagnosis of postoperative tetanus was made and treatment started. Next day, she was not responding to commands. Laboratory reports which were sent previous day, showed a total WBC count of 44,000 cells/mm^3 clinching the diagnosis. It was a case of 'leukaemia'. Neck rigidity was due to leukaemic infiltrates in meninges. Now you know the cause of uterine bleeding!!!

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

Magnesium sulphate can be used to treat autonomic system complications and control of spasms in tetanus. It may be used alone or in combination with benzodiazepines. A loading dose of 5 g over 20 min is followed by infusion, the rate of which is titrated to control of spasm as well as reduction of muscle rigidity. Rigidity should be reduced to a level acceptable to the patient permitting swallowing of saliva, mouth care and limb physiotherapy. Total abolition of muscle rigidity is not required as it may lead to hypotonia. Loss of patellar reflex may not be a valid indicator of hypermagnesaemia in all patients, as the reflex is sometimes masked by rigidity and tends to lost early in elderly patients. The patients, therefore, should be monitored closely for depression of ventilation. Magnesium sulphate may not reduce the need for mechanical ventilation in patients with severe tetanus but may help in reducing the requirement of other sedatives.

MULTIPLE CHOICE QUESTIONS

1. Which of the following statement is false in tetanus?
A. Exotoxins are produced by ***Clostridium tetani***
B. The toxin gets fixed to motor cells of anterior horn cells
C. It stimulates the release of cholinesterase
D. Period of onset is more important than incubation period

2. Favourable conditions for development of tetanus include following *except:*
A. Injury
B. Foreign body
C. Devitalised tissues
D. Aerobic conditions

3. Hyperbaric oxygen can be used in following conditions *except*:
A. Gas gangrene
B. Decompression sickness
C. Carbon monoxide poisoning
D. Necrotising fasciitis

4. Following facts are true in gas gangrene *except*:
A. It is caused by ***Clostridium welchii***
B. Severe myonecrosis is a feature
C. Brownish fluid has foul odour
D. Alfa toxin causes cell membrane damage

5. Positive Nagler reaction in gas gangrene is caused by:
A. Lecithinase
B. Beta toxin
C. Proteinase
D. Hyaluronidase

6. Sponge like consistency of the part is typically seen in which condition:
A. Necrotising fasciitis
B. Diabetic ulcer leg
C. Gas gangrene
D. Pyomyositis

7. Following are true for *Clostridium welchii except*:
A. Anaerobic
B. Gram-positive
C. Nonspore bearing
D. Produces a toxins

8. Following are clinical features of gas gangrene *except*:
A. Crepitus
B. Khaki coloured skin
C. Low grade fever
D. Hypertension

9. Following are features of necrotising fasciitis type II *except:*
A. It is monomicrobial—β haemolytic streptococci
B. It can occur in young healthy individuals also
C. Organism is ***Clostridium perfringens***
D. It can give rise to toxic shock syndrome

10. Which of the following is false to prevent gas gangrene?
A. Antigas gangrene in risk groups
B. Prophylactic antibiotics
C. Blood sugar level should be maintained within 200 mg/dl
D. Avoid tourniquets while operating on crushed wounds in the legs

ANSWERS

1 C 2 D 3 D 4 C 5 A 6 C 7 C 8 D 9 C 10 C

4

Hand and Foot Infections

- Paronychia
- Acute lymphangitis
- Subcutaneous infections
- Terminal pulp space infections
- Apical subungual infection
- Web space infections
- Deep palmar abscess
- Tenosynovitis
- Mycetoma pedis
- Ingrowing toe nail
- What is new?/Recent advances

Introduction

Hand infections are commonly encountered in manual labourers and are precipitated by injury such as thorn prick, cut injuries, etc. In 80–90% of cases, the causative organisms are *Staphylococcus aureus* sensitive to cloxacillin. In remaining cases, *Streptococci*, gram-negative bacilli, anaerobic organisms may also play a role. Irrespective of the site of infection, oedema is commonly encountered on the dorsal aspect because of the following reasons:

- Lymphatics from the palmar aspect of the hand travel through the dorsal aspect to the corresponding lymph node.
- Presence of the loose areolar tissue in the dorsum of the hand.
- Hand infections can be severe in immunocompromised, systemically ill and diabetic patients. It can spread so rapidly to cause septicemia and even death.
- It is unfortunate that in cases of gas gangrene and spreading infection, amputation may have to be done.

PEARLS OF WISDOM

Oedema is the chief cause of stiffness of the fingers. Hence, early physiotherapy should be encouraged.

Classification (Table 4.1)

SUPERFICIAL INFECTIONS

PARONYCHIA[1]

It means near the nail. It is the commonest type of hand infection. There are two types of paronychia, acute and chronic.

Acute paronychia (Fig. 4.1)

- It occurs due to trimming of the nail or ingrowing nail.
- Infection which is subcuticular starts in the lateral sulcus and ***spreads all around*** (paronychia means 'run around'). This is because eponychium (skin overlying the nail base) is adherent to the nail base. Hence, the infection spreads

[1]Paronychia—infection affects the base of nail.

Table 4.1 Classification of hand infections

Superficial infections	Deep infections
1. Paronychia	1. Terminal pulp space infection
2. Subcutaneous infections	2. Apical subungual infection
3. Infection of dorsal space	3. Volar space infection
4. Acute lymphangitis	4. Web space infection
	5. Midpalmar space infection
	6. Tenosynovitis

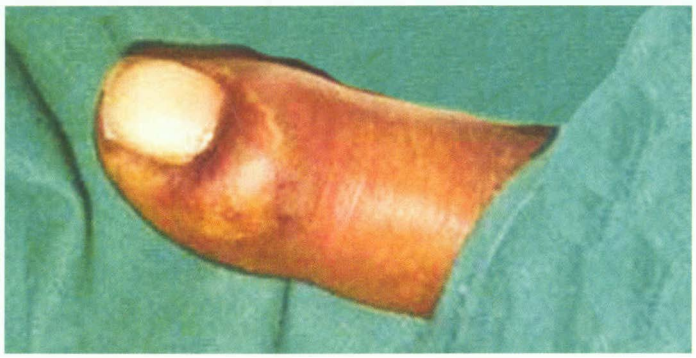
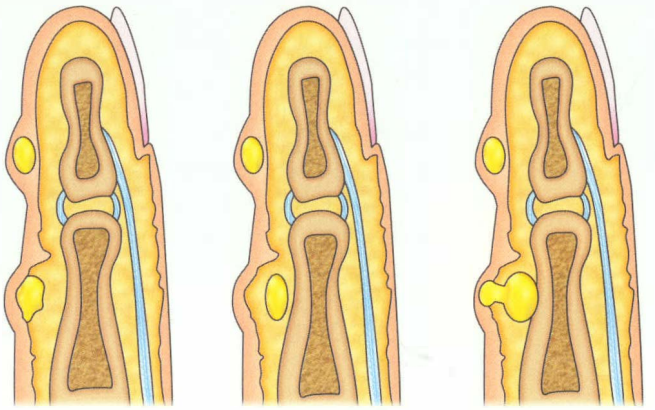


Fig. 4.1: Acute paronychia



Figs 4.2 to 4.4: Intraepidermal abscess, intradermal abscess anc subcutaneous abscess respectively. **Fig. 4.4:** Collar-stud abscess also

beneath the nail base. The affected finger is painful. Throbbing pain suggests presence of pus. Even collection of 0.5 ml pus produces severe pain. Low grade fever may be present.

Treatment

- Early cases (before formation of pus) can be managed by soaking, elevation, antibiotics and immobilisation.
- Using a digital block (with 5 ml of 2% *plain lignocaine* injected into the root of the digit), incision and drainage is done by incising the eponychium. ***Adrenaline should not be used for infiltration*** in the finger, penis and ear lobule as these areas are supplied by end-arteries (no collateral circulation). Adrenaline is a vasoconstrictor and can cause gangrene. Pus is sent for culture and sensitivity. Antibiotics are given. Dressings are applied.

Chronic paronychia[1]

- It is *not due to bacterial infection*. It is due to **fungal infection**—moniliasis or due to candida infections.
- It is common in women who wash the clothes, utensils, etc. and have constantly wet fingers. As a result of this, fungal infection takes place. The infection is insidious in onset, chronic and difficult to eradicate. It produces a dull nagging pain in the fingers. The eponychium is faintly pink and nail is ridged.
- Antifungal agents such as nystatin or tolnaftate solution helps the patient. Rubber gloves should be worn while using hand for washing.

SUBCUTANEOUS INFECTIONS (Figs 4.2 to 4.4)

1. **Intraepidermal abscess:** It is also called purulent blister. Cuts, pricks and burns are also causes of this condition.
2. **Intradermal:** This variety does not produce dome-shaped elevation.
3. **Subcutaneous abscess:** This type of lesion is like that of cellulitis.
4. **Collar-stud abscess:** It results when the epidermal component is connected to dermal component.

Treatment

- Incision and drainage under antibiotic cover. Care should be taken to drain the deeper cavity.

ACUTE LYMPHANGITIS OF HAND

- It is caused by an injury, which may be a minor abrasion.
- The causative organism is ***Streptococcus***.

Clinical features

- Severe pain in the hand with fever, chills and rigors
- Gross oedema of dorsum of hand
- Red, hot streaks over the limb which indicates route of lymphatics.
- Regional lymph nodes are swollen and tender.
  - Infection of little finger—epitrochlear lymph nodes are enlarged.
  - Ring and middle finger—nodes above the clavicle are affected.
  - Index and thumb—axillary nodes are enlarged.

Treatment

- Injection crystalline penicillin 10 lakh units **IV** or **IM** for 5–7 days.
- Higher antibiotics may have to be used depending upon the response.

HERPETIC WHITLOW

- Common in children, may follow herpetic gingivo-stomatitis.
- Most common viral infection of the hand—distal finger.
- It is due to herpes simplex virus (HSV) infection.
- Pain, pruritus, vesicles are characteristics. Fever, lymph-adenitis are present.
- Resolves spontaneously.

[1] Wet nails of women who wash dishes are vulnerable.

DEEP INFECTIONS

INFECTION OF THE TERMINAL PULP SPACE (Felon)

- This space commonly gets infected due to prick injuries which are relatively deep. It is the second common infection of the hand seen in about 25% of the patients.

Anatomy of terminal pulp space

- It is a closed space, formed by fusion of distal flexion skin crease with the deep fascia attached to the periosteum of distal phalanx, just distal to the insertion of flexor digitorum profundus. Each pulp space is subdivided by the presence of numerous septa which pass from deep fascia to the periosteum. Thus 15–20 small compartments are formed (Fig. 4.5).
- The digital artery which is an **end-artery** runs in this closed space (Fig. 4.6)

Clinical features (Key Box 4.1)

- Injury to the affected finger is usually present. Thumb and index are commonly involved.
- Throbbing pain is worse in the dependent position, with nocturnal exacerbations.
- The pulp is indurated, red and tense and is characteristic of this condition.
- Touch, movement, dependent position worsen the pain.

Treatment

- Incision and drainage under digital block—Volar longitudinal incision.

Complications

1. If the pus is not released early, *thrombosis of digital artery* takes place *resulting in osteomyelitis and necrosis of the terminal phalanx* and may result in shortening of the finger (Figs 4.6 and 4.7).
2. Pyogenic arthritis of distal interphalangeal joints.
3. Tenosynovitis secondary to pus requires regular physiotherapy to avoid thickness developing later.
4. Neuroma: It can be painful and can cause discomfort.

KEY BOX 4.1

TERMINAL PULP SPACE INFECTION—FELON

- **F**inger tip pulp abscess
- **E**xtremely painful
- **L**oss of normal resilience of pulp
- **O**steomyelitis of distal phalanx in untreated cases due to thrombosis of digital artery
- **N**o longer recommended **fish mouth** incision

Remember as **FELON**

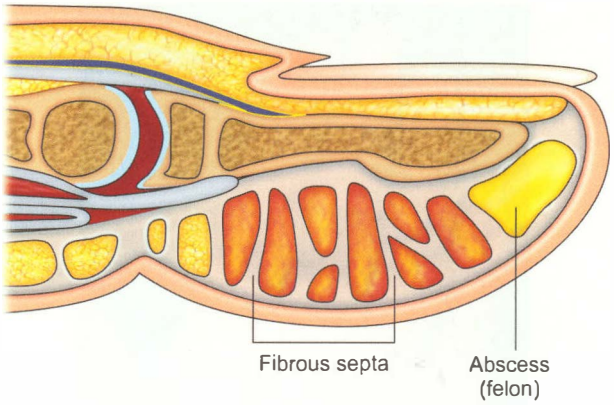


Fig. 4.5: Felon

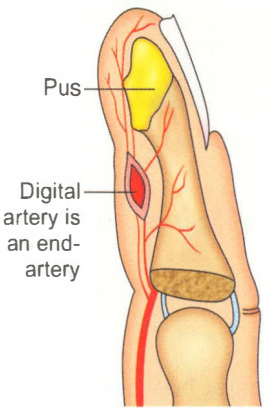


Fig. 4.6: Pulp space infection

Fig. 4.7: Plain radiography of hand showing osteomyelitis of terminal phalanx

APICAL SUBUNGUAL INFECTION

- Infection is confined to the space between the distal quarter of the subungual epithelium and periosteum of the distal phalanx. Penetration by a sharp object is the cause for this condition. It manifests very often as a tender yellow spot beneath the distal portion of the nail. Pain, redness and minimal swelling are the features.
- Tenderness is maximum at the free edge of the nail. Pulp and distal parts of fingers are relatively painless. It is treated by "V" excision of a portion of nail and opening of the abscess cavity with antibiotic cover (Fig. 4.8).

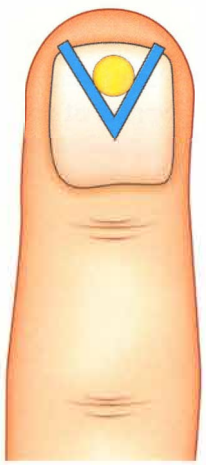


Fig. 4.8: "V" excision of the nail, for apical infection

MIDDLE AND PROXIMAL VOLAR SPACE INFECTIONS (Fig. 4.9)

- These spaces are loose when compared to terminal pulp space. They are filled with fibrofatty tissue.
- Middle volar space is closed but proximally communicates with web space.

Fig. 4.9: Middle and proximal volar space infections

- Swelling is tender and indurated. Finger is held in the position of flexion.
- Treated by transverse incision and drainage of the pus.

WEB SPACE INFECTIONS

- Web spaces are the triangular spaces in between the four divisions of the palmar aponeurosis. *They are 3 in number* (Fig. 4.10). Thumb has no palmar aponeurosis. They are filled with subcutaneous fat and posteriorly covered by metacarpal bones.

Causes of web space infection

- Penetrating injuries
- Spread of proximal volar space (palmar space) infection.
- Lumbrical canal infection—suppurating tenosynovitis.

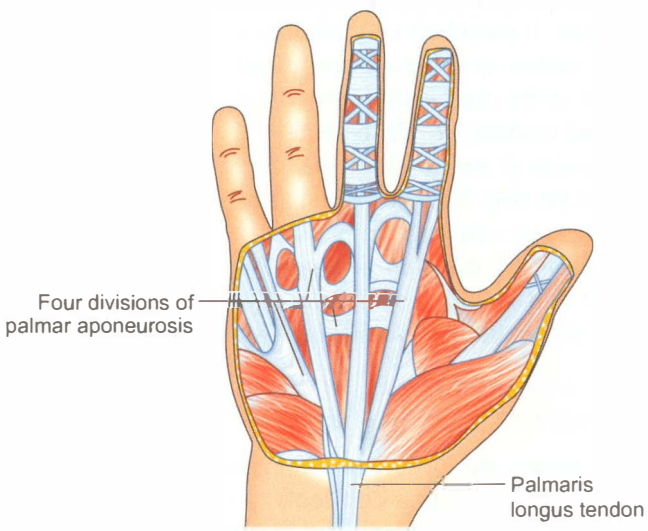


Fig. 4.10: Three web spaces

Clinical features

- Pain and swelling of palm in the region of web space.
- Extremely tender and hot swelling.
- **Finger separation sign:** Adjacent fingers are separated du to oedema (Key Box 4.2).
- Gross oedema of the dorsum of hand.
- If untreated, pus from one web space can spread to the othe web space and to the other proximal volar space.

KEY BOX 4.2

WEB SPACE INFECTIONS

- 3 web spaces
- Finger separation sign
- Gross oedema of dorsum
- Spread to other web space

Treatment

Under anaesthesia, a transverse skin incision is made and the pus is drained (Fig. 4.11). The cavity is treated like any other abscess cavity. The skin edge is trimmed in such a way as to leave a *diamond-shaped* opening to get better drainage.

DEEP PALMAR ABSCESS

- Infection of midpalmar space results in deep palmar abscess.
- Midpalmar space is the space behind the palmar aponeurosis and in front of the metacarpal bones.
- Since palmar fascia is thick, strong and unyielding, pus collects deep to palmar fascia. If it is due to penetrating injuries, it collects in the subcutaneous plane like collar-stud abscess. In the centre of palm, there is no subcutaneous tissue. Hence, pus collects beneath the thick dermis.

Surgical anatomy of palmar fascia and aponeurosis (Fig. 4.12)

- The central thick fibrous part of palmar fascia is palmar aponeurosis.

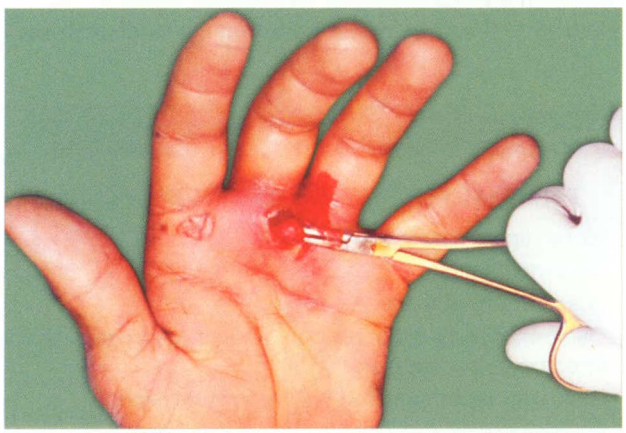


Fig. 4.11: Drainage of web space abscess

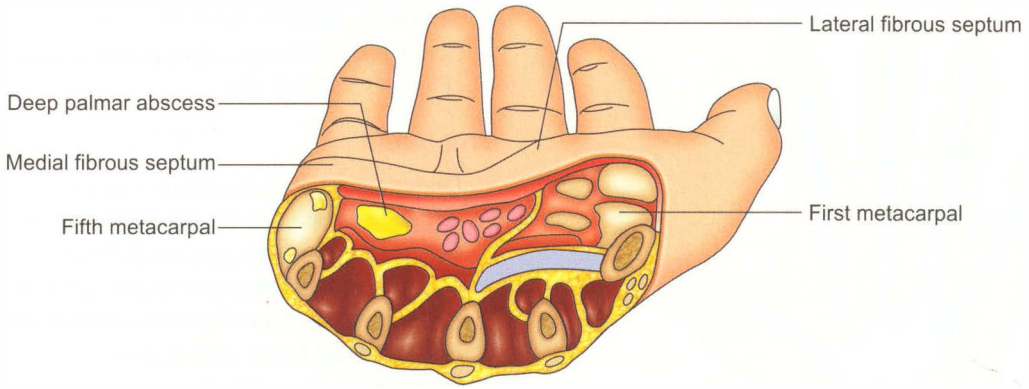


Fig. 4.12: Cross-section showing deep palmar space

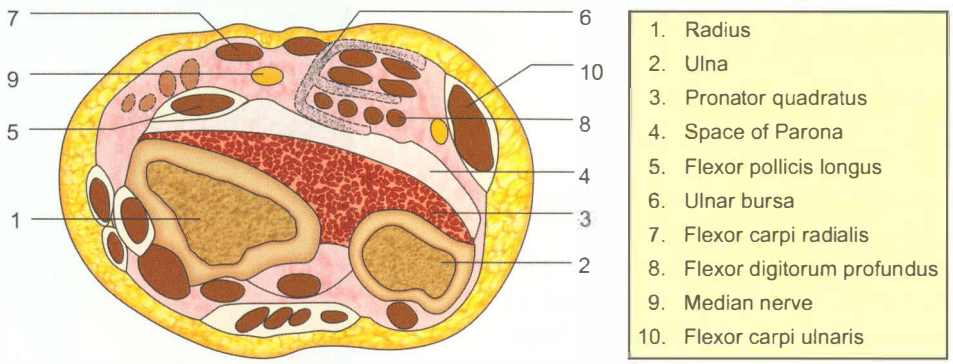


Fig. 4.13: Space of Parona and it's relationship to other structures

- Palmar fascia covers long flexors.
- Apex of the triangular palmar aponeurosis is continuous with flexor retinaculum and palmaris longus tendon.
- Distally, it forms 4 longitudinal digital bands and attaches to bases of proximal phalanges.
- Two fibrous septa extend from medial and lateral margins of palmar aponeurosis. They are medial and lateral fibrous septa. The septum is attached to 5th and 3rd metacarpals respectively.
- Deeper to flexor tendons, digital arteries and nerves lies the midpalmar space.
- The midpalmar space is continuous with anterior compartment of the forearm *via* carpal tunnel. This space is called '**space of Parona**' (Fig. 4.13).

Source of infection

- Penetrating injuries
- Haematoma
- Suppurative tenosynovitis

Clinical features

- Obliteration of *normal concavity of the palm*
- *Gross oedema* of the dorsum of the hand
- *Extreme tenderness* in midpalmar space
- Fingers are held in *flexion* at the *metacarpophalangeal* ***(MP)*** joint because the palmar aponeurosis gets relaxed in this position. ***MP*** *joint movements are painful.*
- IP (interphalangeal) joint movements are not painful.

PEARLS OF WISDOM

Thus, swollen palm, oedema of the dorsum of the hand, flexed attitude of MP joint and separated fingers give the picture of a frog hand.

Treatment

- Under anaesthesia, a transverse crease incision is made and once the palmar aponeurosis is seen, it is split *longitudinally* in the direction of the fibres *to avoid damage to nerves and vessels.*
- Abscess cavity is treated in the usual manner.

ACUTE SUPPURATING TENOSYNOVITIS[1]

Surgical anatomy of flexor tendon sheath arrangements

- The flexor tendon sheaths which enclose the tendons run along the whole length of the finger. In the palm, the medial tendons

[1]Henry Hamilton Bailey lost his left index finger because of acute tenosynovitis.

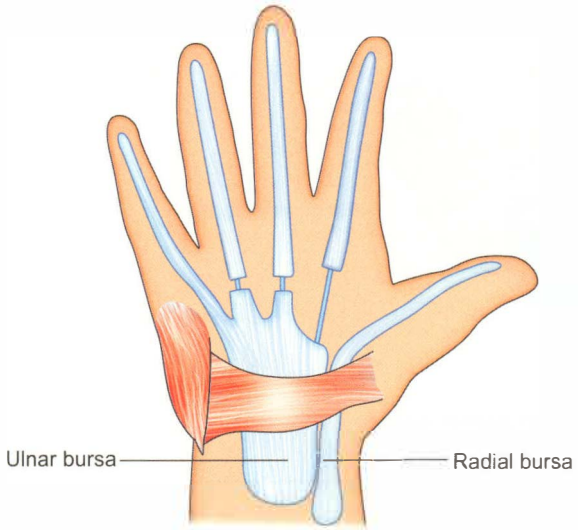


Fig. 4.14: Flexor tendon sheath arrangements

are enclosed by a common synovial pocket called "ulnar bursa" and on the lateral side by "radial bursa" (Fig. 4.14).

- These two are communicating in 80% of the cases. In 25% of cases, the flexor tendon sheath of the thumb is communicating with radial bursa and of the little finger with ulnar bursa.
- Thus, infection of the fingers (flexor tendon sheath) can involve the entire hand.
- The synovial sheaths of flexor tendons extend from the bases of the terminal phalanges to the heads of the metacarpal bones.

Clinical features (Key Box 4.3)

- The patient gives history of pricking type of injuries.
- Symmetrical, fusiform painful enlargement of finger
- Flexed, fixed finger—***'Hook sign'***
- ***IP joint movements are very painful:*** Severe pain on passive finger extension.
- ***MP joint movements are not painful:*** This sign differentiates suppurating tenosynovitis from deep palmar abscess.
- When there is infection of ulnar bursa, the maximum tender spot is in between the two palmar creases. This sign is described as ***'Kanavel's sign'***.

KEY BOX 4.3

SUPPURATING TENOSYNOVITIS

- Sharp prick injuries
- Hook sign—bent finger
- IP joint movements painful
- MP joint movements need not be painful
- **'Kanavel's sign'**—ulnar bursa infection

- Similarly, there is tenderness over lateral side, over th flexor pollicis longus sheath when radial bursa is involve (Figs 4.15 and 4.16).

Treatment

- Under anaesthesia, multiple incisions may have to be give to decompress the flexor tendon sheaths, so as to relieve tension to drain the pus, exudate, etc.
- The cavity is irrigated with antiseptic solution.
- In severe cases a small plastic catheter should be introduce into synovial bursa and it should exit by counter incisio in the palm for antibiotic irrigation.

Fig. 4.15: Suppurating tenosynovitis

Fig. 4.16: Grossly swollen hand in a diabetic patient. It was looking dangerous, but responded to conservative line of treatment

- Postoperatively, appropriate antibiotics are given for about 2 weeks.
- Hand is positioned in elevated position to reduce oedema.

Complications

1. Stiffness of the fingers
2. Suppurating arthritis of the joints
3. Osteomyelitis
4. Loss of tendon, digit
5. Spread of infection to ***space of Parona***. It is the space deep to the flexor profundus and superficial to pronator quadratus in the lower end of forearm. Patients present with swelling of the forearm along with gross oedema of the hand.
   In addition to the treatment mentioned above, a separate incision may have to be given in the lower forearm for a better drainage of the pus.

GENERAL PRINCIPLES AND MANAGEMENT OF HAND INFECTION

- Early diagnosis, early splinting and elevation.
- Early proper drainage
- Proper incision—preferably a crease incision
- **Elevation** of hand to reduce oedema
- Pus culture and sensitivity
- Cloxacillin 500 mg, 6th hourly for 7–10 days, with metronidazole 400mg, 8th hourly for 7–10 days. Otherwise, higher antibiotics such as cephalosporins may have to be given.
- Physiotherapy to decrease the stiffness of the fingers.
- Tetanus prophylaxis in high-risk patients.

Recommended antibiotics

• *Staphylococcus aureus*	First generation cephalosporins
• Anaerobics / *Escherichia coli*	Clindamycin or β-lactamase as inhibitors, amoxicillin clavulanate potassium
• Herpetic whitlow	Antivirals

Atypical Mycobacterial infections

- Affects tendon sheaths
- Causes swelling and stiffness
- Pain and redness
- Exploration, excision of infected lining of tendon sheaths will give diagnosis
- Mycobacterium marinum is the common bacteria

Position of function

The hand is held as if holding a cup or a glass

- Extension at wrist: 25 degrees
- Flexion at metacarpophalangeal joint 60 degrees
- Flexion at interphalangeal joint 10 degrees

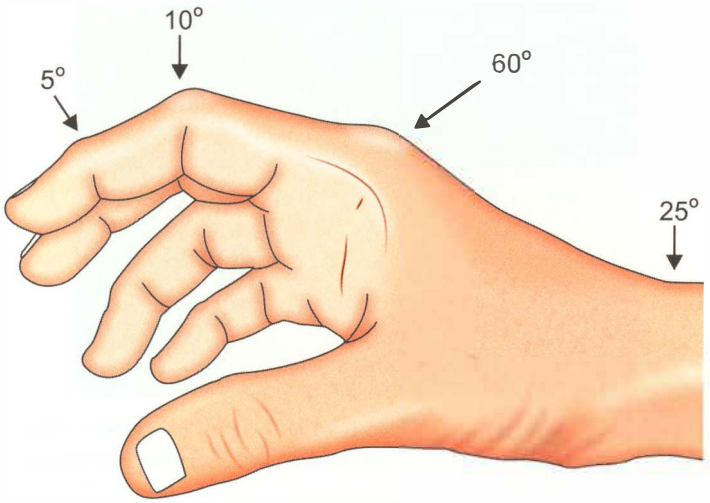


Fig. 4.17: Position of function

- Flexion at distal interphalangeal joint 5 degrees
  Thumb in alignment with forearm (Fig. 4.17)

OTHER HAND INFECTIONS

1. Compound palmar ganglion (*see* page 238)
2. Barber's pilonidal sinus (inter digital)
3. Madura mycosis (next page)
4. ***Orf virus:*** Highly contagious pustular dermatitis due to parapox virus infection.
5. ***Milker's nodes:*** It is a viral disease transmitted by handling Cow's udder.
6. ***Human bites:*** Common organism is *Staphylococcus*. The wound is explored and proper treatment given.

FOOT INFECTIONS

MYCETOMA PEDIS

- It refers to chronic granulomatous lesions of the foot involving skin and subcutaneous tissues. The disease not only involves skin and subcutaneous tissue but also the deeper structures such as bones resulting in osteomyelitis.
- **Gross thickening of subcutaneous tissue results in convexity of the instep of the foot** that is characteristic of this condition (Fig. 4.18). Chronic suppuration, **multiple sinuses**, sulphur granules in the discharge are characteristic of mycetoma pedis (Fig. 4.19).
- Bare foot walking which results in repeated minor trauma, implants the organisms within subcutaneous tissue. It starts as a **pale, painless single nodule**. Later multiple nodules develop and rupture resulting in multiple sinuses.

Fig. 4.18: Flattening of the foot (*Courtesy:* Prof Madakatti, HOD Surgery, Karnataka Institute of Medical Sciences, Hubli, Karnataka)

Fig. 4.19: Mycetoma pedis

Types

1. **Bacterial mycetoma:** It is due to *Nocardia madurae* or due to actinomyces. These organisms are normally present in the soil.
2. **Fungal mycetoma:** It is caused by *Madurella mycetoma*, etc.

Diagnosis

- Sulphur granules in the discharge, plain radiograph of the foot (Fig. 4.20).

Treatment

1. Broad-spectrum antibiotics to treat secondary infection along with **dapsone 100 mg, twice daily is the choice**. Treatment may have to be continued for 1–2 years.
2. Fungal mycetoma may not respond to antibiotics.
3. Amputation may be necessary, in refractory cases, to get rid of a deformed, diseased limb.

INGROWING TOE NAIL (Onychocryptosis)

- It is also described as embedded toe nail. Exact aetiology is not clear. However, a few patients have family history of this condition. Trimming the nail too much may result in ingrowing toe nail.

Fig. 4.20: Plain radiograph of the foot showing extensive rarefaction of the bones

- As the nail grows inside, some degree of infection sets in, resulting in development of granulation tissue which starts pouting. The condition is painful, disturbing and unsightly.

Treatment

- **Conservative:** Dressing with antiseptic agents such as iodine. Copper sulphate can be applied to treat extra granulation tissue. Appropriate antibiotics are given.
- **Surgical:** Under local anaesthesia, a portion of the involved nail upto the base is removed followed by application of phenol to the growing point of the nailbed at its base. It takes about 10–15 days for complete healing.
- **Zadik's or Fowler's operation:** The principle of this radical procedure is to expose the lateral spike and germinal matrix. This is achieved by incising the skin in lateral margin and root of the nail.
- Hand infections—see the photographs in the next page (Figs 4.21 to 4.25).

MISCELLANEOUS

Pyogenic granuloma (Fig. 4.26)

- It is not a true granuloma but it is a capillary haemangioma
- It is due to trauma not due to infection (pyogenic is a misnomer)
- It is smooth, red, lobulated
- Can be painful
- They bleed due to minor truama
- In fingers, persistent irritation results in excessive granulation tissue
- It can occurs in oral cavity also
- It is also called pregnancy tumour

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- I have added a new Key Box—FELON
- Surgical anatomy of palmar fascia and aponeurosis has been added
- All topics are updated
- Pyogenic granuloma has been added

HAND INFECTIONS—A FEW PHOTOGRAPHS

Figs 4.21 and 4.22: Very severe dorsal space infection in a diabetic patient. Observe the constriction effect caused by bangle. They need to be removed in all cases of hand infections

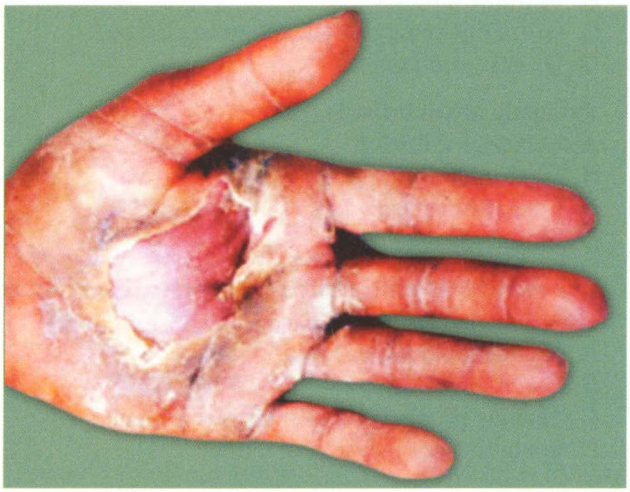


Fig. 4.23: Healing palmar space infection. Observe the cuticle being raised

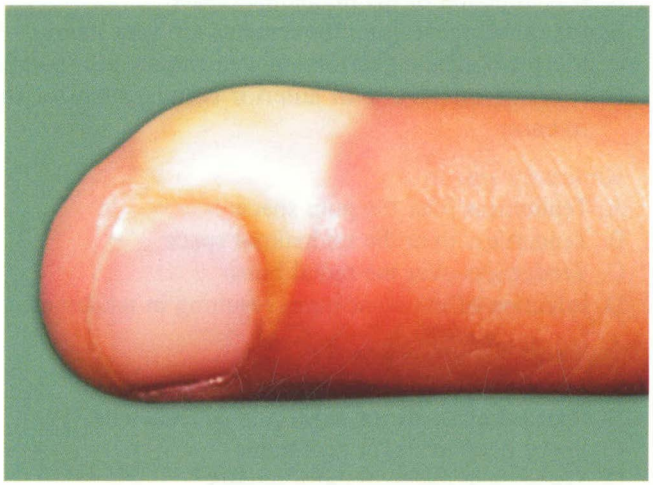


Fig. 4.24: Acute paronychia—very painful condition, can be treated very easily

Fig. 4.25: Wet gangrene of the hand (*Courtesy:* Dr Maruthu Pandyan, Government Medical Collage, Madurai)

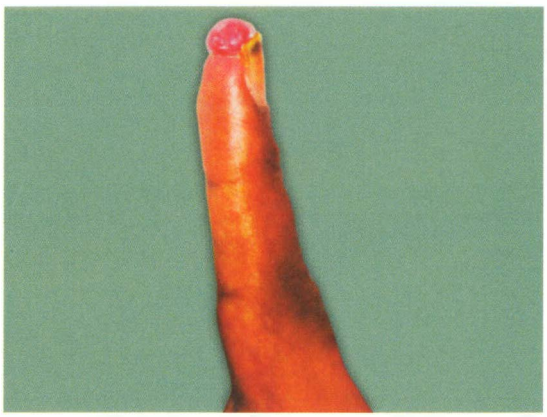


Fig. 4.26: Pyogenic granuloma

(All the photographs in this chapter are contributed by Dr P Sreenivasa, Postgraduate student (2000–2003) and Prof Mahimanjan Singh, Head of the Department of Surgery, Govt Medical College, Mysore, Karnataka)

MULTIPLE CHOICE QUESTIONS

1. Which of the following statement is false in paronychia?
A. Chronic paronychia is caused by fungal infections
B. It is a deep infection
C. It is the commonest hand infection
D. Infection spreads beneath the eponychium

2. Following are true for terminal pulp space infection *except*:
A. It is called Felon
B. It may cause digital arterial thrombosis
C. Osteomyelitis of distal phalanx will not occur due to extensive collaterals
D. Pyogenic arthritis can occur

3. Following are true for deep palmar abscess *except*:
A. It is infection of the midpalmar space
B. Interphalangeal joint movements are very painful
C. Metacarpophalangeal joint movements are very painful
D. It communicates with anterior compartment of the forearm

4. Kanavel's sign is:
A. Tenderness over the pulp space
B. Tenderness over the midpalmar space
C. Tenderness over the radial bursa
D. Infection of the ulnar bursa resulting in tenderness

5. Space of Parona refers to:
A. Space deep to palmar aponeurosis
B. Space deep to flexor digitorum superficialis
C. Space deep to flexor profundus
D. Space deep to flexor carpi ulnaris

6. Following is true for acute suppurating tenosynovitis *except:*
A. Finger is fixed and flexed
B. Interphalangeal joint movements are not painful
C. Metacarpophalangeal joint movements may be painful
D. Kanavel's sign may be positive

7. Characteristic feature of mycetoma pedis is:
A. Disease affects bones
B. Starts as a pale painless single nodule
C. Extensive rarefaction of the bone
D. Convexity of the instep of the foot

8. Following are clinical features of mycetoma pedis *except:*
A. Crepitus
B. Multiple sinuses
C. Gross thickening of subcutaneous tissues
D. Disease is chronic

9. Pyogenic granuloma is due to:
A. Bacterial infection
B. Viral infection
C. Fungal infection
D. Trauma

10. Following are the principles of treatment of hand infections *except:*
A. Hand should be elevated
B. Early splinting
C. Tetanus prophylaxis in high-risk patient
D. Pencillins are the drug of choice

ANSWERS

1 B 2 C 3 B 4 D 5 C 6 B 7 D 8 A 9 D 10 D

5

Chronic Infectious Disease

- Actinomycosis
- Leprosy
- Deformities in leprosy
- Syphilis
- AIDS
- What is new?/Recent advances

Introduction

Actinomycosis, leprosy, syphilis and AIDS are the chronic diseases discussed in this chapter. Actinomycosis is a rare disease, leprosy is of more interest to the skin specialists and AIDS is an interesting topic to all clinicians. Hence, only relevant aspects of each of these diseases as far as general surgeons are concerned are being discussed here.

ACTINOMYCOSIS

Actinomycosis is caused by *Actinomyces israelii*, an anaerobic, gram-positive branching filamentous organism (ray fungus). Normally present in the oral cavity, tonsillar crypts and dental cavities, they become pathogenic in the presence of trauma. Three types of actinomycosis which are of some interest to general surgeons have been discussed here.

FACIOCERVICAL ACTINOMYCOSIS

- It is common in patients with poor oral hygiene, bad caries tooth, etc.
- The organisms produce a subacute or chronic inflammation for many months to years and produce lumpy jaw (Key Box 5.1 for differential diagnosis).
- Eventually cheek, mandible, jaws and salivary glands are involved resulting in suppuration.

Clinical features

- **Extensive induration** (marked induration) of lower jaw (mandible) and gums gives consistency of bone.
- **Multiple subcutaneous nodules** over bluish-coloured skin of the jaw.
- The nodules rupture resulting in ***multiple discharging sinuses***.
- The discharge contains ***sulphur granules*** which are gram-positive mycelia surrounded by gram-negative clubs.
- **Lymph nodes are not involved.**

ACTINOMYCOSIS OF THORAX AND LUNG

- It is common in children, caused by inhalation of ray fungus.
- Over a period of years, it produces actinomycosis of lung with involvement of pleura. Later it involves the chest wall, resulting in multiple discharging sinuses.
- There may be associated empyema and can easily spread to liver.

ACTINOMYCOSIS OF THE RIGHT ILIAC FOSSA AND LIVER

- It commonly occurs after surgery when there is **mucosal injury** or discontinuity, e.g. after appendicectomy.

KEY BOX 5.1

DIFFERENTIAL DIAGNOSIS

- Jaw tumour
- Osteomyelitis of jaw
- Malignancy of oral cavity

- The organisms which are normally present in the gut slowly migrate into pericaecal tissue, then into the soft tissue, subcutaneous tissue, and produce subacute or chronic low grade inflammation.
- No compromise with bowel lumen.
- Once the portal venous radical gets involved, spread to the liver occurs.

Clinical features

- History of appendicectomy is present in almost all cases.
- 3–6 months later, fever and swelling in right iliac fossa appears. Fever is probably due to pyaemia.
- On examination, there is a mass in the right iliac fossa which is indurated, nodular and fixed.
- Late stages produce multiple discharging sinuses, sometimes faecal matter and sulphur granules. Unlike tuberculosis, the lymph nodes are not enlarged.

Differential diagnosis

- Carcinoma caecum, Crohn's ileocolitis, pericolic abscess, etc.

Treatment of actinomycosis in general

- It is low grade chronic disease, difficult to eradicate.
- Inj. crystalline penicillin 10 lakh units once a day for 6–12 months. Tetracycline and lincomycin are the other alternatives.
- Sinuses in the jaw may have to be excised and osteomyelitis has to be curetted out.
- Actinomycosis of the right iliac fossa may need right hemicolectomy.

LEPROSY (HANSEN'S DISEASE)

- Leprosy is caused by *Mycobacterium leprae*, an acid-fast bacillus. Poverty, poor hygiene and population (overcrowding) facilitate the spread of the disease.
- The disease is contracted in childhood or adolescence, but it manifests after a latent period of 2–5 years.
- **Nasal secretions** are the main source of infection but ***active ulcers, sweat*** also contain lepra bacilli.
- Leprosy predominantly affects **skin, upper respiratory tract (nasal cavity) and nerves**. Thus, characteristic lesions of leprosy include an anaesthetic patch of skin, thickened nerves, a deformed leonine face and collapsed nose.

Types (Table 5.1)

1. **Tuberculoid leprosy:** It occurs in patients with good immunity with strong tissue response.
2. **Lepromatous leprosy:** It occurs in patients with poor immunity with poor tissue response.
3. **Borderline leprosy:** It can be borderline lepromatous or borderline tuberculoid leprosy depending upon the immune response.

Treatment

1. **Lepromatous and borderline lepromatous leprosy (Multibacillary disease)**
   - 3-drug regimen is the most ideal treatment.

   Dapsone 100 mg/day,
   Clofazimine 50 mg/day
   Rifampicin 600 mg once monthly, supervised.
   Clofazimine 300 mg once monthly, supervised.
   } For a minimum period of 2 years. Skin smear should be negative.
2. **Tuberculoid and borderline tuberculoid leprosy (Paucibacillary disease)**
   - Dapsone 100 mg daily.
   - Rifampicin 600 mg once a month, supervised.

   } For a period of 6 months.

DEFORMITIES IN LEPROSY

I. Primary deformity (Figs 5.1 to 5.3)

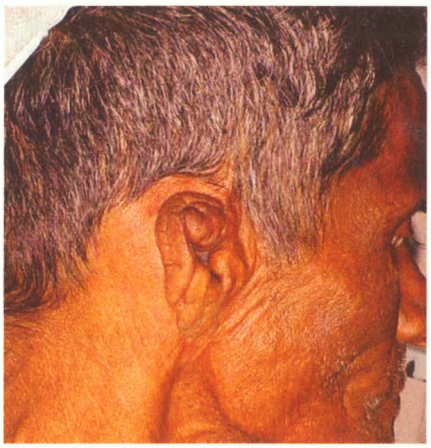
It occurs directly due to the disease.

- **Face:** It is involved in lepromatous leprosy and is described as leonine facies with multiple nodules over the face, pigmentation, *loss of lateral portion* of the eyebrows (madarosis), collapse of bridge of the nose due to destruction

Table 5.1 Comparison of tuberculoid leprosy with lepromatous leprosy

	Tuberculoid leprosy	Lepromatous leprosy
1. Cell-mediated immunity	Strong	Low
2. Histology	Giant cells, epithelioid cells, histiocytes, lymphocytes are present. Bacilli are few.	Bacilli distending the macrophages—'GLOBI'. Plenty of bacilli invading nerves, adnexa, sweat glands, etc.
3. Clinical	Localised. Anaesthetic, hypopigmented, raised skin patch. Early nerve damage, nerve thickening is a characteristic feature. Involvement of face and nose is not seen.	Generalised. Erythematous multiple macular rashes. Nerve involvement is usually not seen. Leonine facies, collapse of the bridge of the nose are characteristic.
4. Prognosis	Good	Not good.

Fig. 5.1: Observe small muscle wasting in both the hands



Leonine facies, collapse of the bridge of the nose are a few other features of lepromatous leprosy

Fig. 5.3: Observe deformed ear

Fig. 5.2: Autoamputation of the toes in a case of leprosy

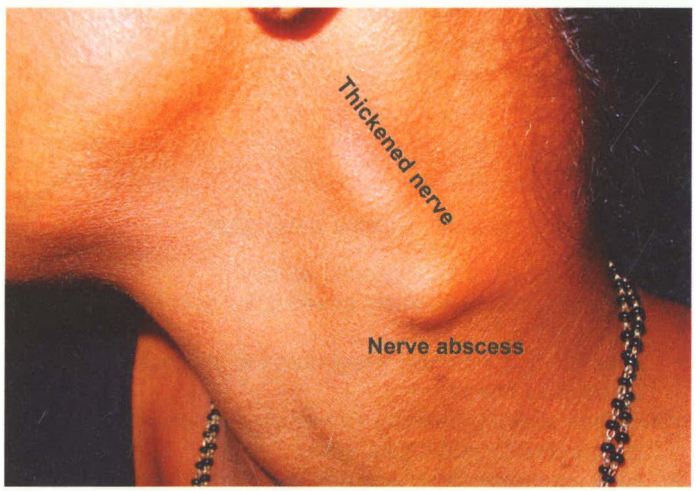


Fig. 5.4: Nerve abscess and thickened postauricular nerve

of nasal cartilages (warm and moist area) and paralysis of facial nerve.

- **Hands:** Involvement of ulnar nerve at the elbow and median nerve at wrist gives rise to ***'claw hand'***.
- **Foot:** Posterior tibial nerve is involved at the ankle leading to clawing of the toes. Foot drop occurs when lateral popliteal nerve below the knee joint is involved.

Ulnar nerve abscess and small muscles atrophy in the hand (Figs 5.5 and 5.6 and Key Box 5.2)

CLINICAL NOTES

MS exam case, KMC, Manipal (2004)

A 28-year-old lady complained of swelling in the posterior triangle of 8 months duration. It was a bit irregular with restricted mobility and it was tender. 3 candidates offered nonspecific lymphadenitis as a diagnosis. Only one candidate thought of nerve abscess. The clue was thickened nerve above. It was a case of Hansen's disease (Fig. 5.4).

Fig. 5.5: Small muscles atrophy in the hand

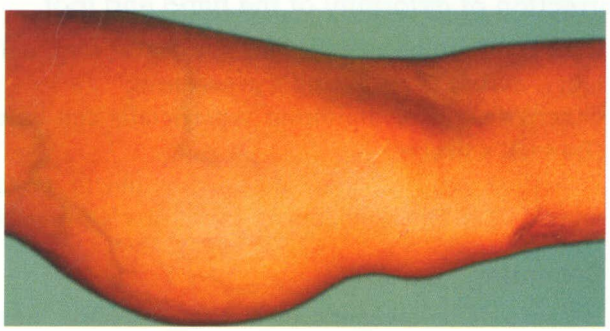


Fig. 5.6: Ulnar nerve abscess

CLINICAL NOTES

A young man, 33 years of age, presented to the outpatient department with inability to use his right hand for the last 6 months. He said he could not work, or hold objects. Examination revealed atrophy of small muscles, specially hypothenar muscles and interossei muscles. A 2 cm 'skin ulcer' was also noticed. On careful questioning, he said he also had loss of sensation. Further examination revealed a 4 cm × 3 cm oval swelling on the medial side of right arm. It was firm. Some fluctuation could be elicited—

What is the diagnosis?

*Ulnar nerve abscess (cold)**

**A case of leprosy*

This is a case of pure neuritic leprosy with nerve abscess usually caused by *Mycobacterium leprae*. It is an uncommon manifestation. No other peripheral nerve was involved. Differential diagnosis will be ulnar nerve schwannoma.

KEY BOX 5.2

ULNAR NERVE ABSCESS

- May be a part of pure neuritic leprosy
- Presents as oval fluctuant swelling on the medial side of arm
- Granuloma is common. Progression to abscess occurs in tuberculoid leprosy.
- **Schwann cell** is affected. Slowly whole endoneurial zone is occupied by endothelial cells.
- High resolution ultrasound can demonstrate echotexture of masses.
- MRI—with the post-gadolinium T_1W sequence—peripheral rim enhancement with central necrosis.
- Treatment: Incision, drainage, excision of granulomatous mass followed by treatment for leprosy.

Correction of deformity of the face by plastic reconstruction

1. A prosthesis to correct the nose
2. Lateral tarsorrhaphy, to prevent exposure keratitis.
3. Temporalis muscle flap to the upper eyelid to prevent exposure keratitis.

Correction of deformity of the hand and foot

- Claw hand can be corrected using extensor carpi radialis brevis muscle **(Paul Brand's procedure)**.
- Otherwise, flexor digitorum superficialis can be used **(Bunnell's procedure)**.
- Foot drop can be corrected by using tibialis posterior muscle tendon transfer **(Ober's and Barr's procedure)**.

II. Secondary deformity

- Because of involvement of the nerves, sensations are impaired or lost. As a result of this, ulcers on the fingers, a deep penetrating, *perforating* ulcer over sole of the foot and even autoamputation of toes can occur.

Treatment of secondary deformity

Nonhealing ulcer over the sole of foot is corrected by applying a POP (plaster of Paris) posterior slab. It takes off the pressure and thus the ulcer heals. If calcaneus is involved due to osteomyelitis, the bone has to be curetted out followed by regular dressings.

SYPHILIS: FRENCH DISEASE, GREAT POX

- This is a sexually transmitted disease caused by *Treponema pallidum*. It is a delicate spiral organism (spirochaete).
- Syphilis is infective only in its early stage. Early lesions are predominantly situated in moist areas such as genitalia and oral cavity.

Clinical presentation

- *See* Key Box 5.3 for congenital syphilis

I. Early syphilis
II. Late syphilis

I. Early syphilis

1. **Primary syphilis:** Classically, a genital chancre occurs in the penis or vulva after 3–4 weeks of sexual exposure.
   - This chancre is shallow, indurated, painless ulcer called Hunterian chancre. Associated inguinal nodes which are shotty, multiple, nontender clinch the diagnosis (Fig. 5.7).
   - Extragenital chancres can occur in the lips, tongue, nipple, etc. They produce large enlargement of the corresponding lymph nodes. Chancres in the rectum and perianal region are common in homosexuals. They are painful and resemble anal fissures.

Investigations

Serological tests for syphilis

A. Nonspecific: VDRL, Kahn, Meinicke, Wasserman

B. Specific treponemal antigen tests

- CFT—Complement fixation test

KEY BOX 5.3

CONGENITAL SYPHILIS

Early

- Snuffles (rhinitis), epiphysitis, periostitis, osteochondritis.

Late

- **HUTCHINSON'S TRIAD**
  - Interstitial keratitis
  - 8th nerve deafness
  - Hutchinson's teeth

Fig. 5.7: Genital chancres

- TPHA—TP haemagglutination test
- TPI—TP immobilisation test
- FTA-Abs—Fluorescent treponema antibody absorption test
- Demonstration of *Treponema pallidum* in the clear exudate from the lesion by dark field microscopy confirms the diagnosis.

2. **Secondary syphilis:** It appears after 6–12 weeks of spirochaetaemia.
   - It is characterised by ***bilateral, symmetrical, coppery red rashes*** which are generalised.
     The *rash* is macular or papular, *never vesicular*. Papules on moist sites such as vulva and perineum enlarge to form condylomata lata—fleshy wart-like growths.
   - Small superficial ulcers in the mouth join to form ***snail track ulcers.***
   - Generalised lymphadenopathy involving epitrochlear and occipital nodes can occur.
   - Moth-eaten alopecia, iritis, bone and joint pains.
3. **Latent syphilis:** If secondary syphilis is not treated, it will develop into latent syphilis. There are no signs but serum tests are positive.

II. Late syphilis

It is also called tertiary syphilis. It basically affects vessels causing inflammatory reactions and the end result is as follows: 'Endarteritis obliterans'→ tissue necrosis → ulcers or fibrosis.

This stage develops after 5–15 years of primary syphilis. It causes neurosyphilis and cardiovascular syphilis. Lesser form, a benign lesion is called ***gumma***. Gumma is a syphilitic hypersensitivity reaction consisting of granuloma with central necrosis and sloughing.

Clinical features of gumma

- Typically it is a subcutaneous swelling.
- Affects midline of the body, e.g. posterior 1/3 tongue, sternum, over the sternoclavicular joint.
- Edges are ***punched out*** when the gumma ulcerates.
- Floor contains ***wash leather slough***.
- On healing, it leaves a **silvery, tissue paper scar** (thin scar).

PEARLS OF WISDOM

Gumma can also involve bone, testis and liver (ovary is not involved).

Treatment

1. Primary and secondary syphilis are treated by injection procaine penicillin 10 lakh units IM × 14 days.
2. *In late syphilis:* Treatment is continued for a period of 21 days. With the current effective treatment of syphilis, it is highly unusual to find late cases now.

AIDS AND THE GENERAL SURGEON

- **A**cquired **I**mmuno**d**eficiency **S**yndrome (**AIDS**) is the end stage of a progressive state of immunodeficiency.
  Causative organism: *Human immunodeficiency virus* (HIV).
- The details regarding aetiopathogenesis and immunology about AIDS are discussed in medicine books. **Topics of surgeon's interest are discussed below.**

Prophylactic measures to be adopted by surgeons (healthcare workers) while treating AIDS patients (universal precautions) (Key Box 5.4)

I. In the outpatient department (OPD)

- Gloves[1] must be worn when examining any patient with open wound.
- Gloves should be worn during proctoscopy or sigmoidoscopy.
- Hand gloves and eye protection during flexible endoscopy.
- Use disposable instruments.
- Reusable instruments such as endoscopes are cleaned with soap and water and immersed in glutaraldehyde.
- No surgical procedure involving sharp instruments is performed in the OPD.

II. In the operation theatre

- Operating table is covered with a single sheet of polythene.
- The number of theatre personnel is reduced to minimum.
- Staff with abrasions or lacerations on their hands are not allowed inside the theatre.
- Staff who enter the theatre must wear shoe covers, gloves, disposable, water-resistant gowns and eye protection.

[1]Gloves were introduced by William Halsted to protect his nurse's hands from the harmful effects of carbolic acid (the nurse became his wife).

KEY BOX 5.4

SUMMARY OF UNIVERSAL PRECAUTIONS TO BE TAKEN IN OT

Face mask to be worn
Avoid splashing on body: Be slow
Cover feet and legs: Shoe covers, boot
Extra glove: Double glove
Meticulous technique: Haemostasis, gentleness
Allow only required people
Spectacles: Protects from splash
Kidney tray: Use it to transfer sharps
Remember as **FACE MASK**

- **Double gloves and eye protection** by staff directly involved with the operation (surgeon, assistant, scrub nurse) (Figs 5.8 to 5.10).
- **Surgical technique**
  1. Avoid 'sharps' and needlestick injuries
  2. Prefer scissors or diathermy, to the scalpel
  3. Use **skin clips rather than skin sutures**
  4. Proper autoclaving at the end of surgery
  5. In the event of needlestick injury, allow the part to bleed and wash the part thoroughly in tap water.
  6. AZT—zidovudine, lamivudine and indinavir should be given to health workers following exposure of susceptible areas to infected material from AIDS patients.

Range of surgery in HIV-positive patients

I. Anorectal disease is the most frequent reason for surgical treatment in HIV-positive patients. This is common in homosexuals in whom AIDS is also common. They have been grouped together as "***AIDS anus syndrome***". **Anorectal disease can be classified** into:

- Anal warts, diarrhoea
- Perianal sepsis—abscesses
- Anal ulceration, fissures
- Reduced sphincter tone and anal incontinence in homosexuals.

II. Abdominal pain: This is due to gastrointestinal opportunistic infection usually caused by *Cytomegalovirus* (CMV). It is a type of colitis and produces abdominal pain, cramps, loose stools, blood and mucus in the stools resulting in emaciation. Flexible sigmoidoscopy can reveal severe proctitis due to cytomegalovirus infection. Severe colitis may lead to acute toxic dilatation of colon.

- Biliary tract infection by *Cryptosporidium* can cause **acute cholecystitis**. AIDS-related sclerosing cholangitis can occur resulting in right upper quadrant pain.
- **Abdominal lymphoma** with involvement of liver and spleen also causes abdominal pain.
- Severe abdominal pain, may be due to **perforation** of small or large bowel which should be treated as an emergency.
- **Appendicitis** is also common due to CMV infection.

III. Lymphomas and Kaposi's sarcoma, etc. They are due to reduced cellular immunity due to following reasons:
1. Immunosuppression used in organ transplantation
2. Severe malnourishment

CLINICAL NOTES

A 23-year-old-lady, carrying a 6-month-old child, presented to us with severe dysphagia and odynophagia. Endoscopy revealed extensive, unusual oesophageal ulcers which prompted us to conduct HIV test. It was positive. We called her husband and wanted to convey the message to him. Before we conveyed, he said, 'I am HIV-positive, my wife is HIV-positive and even my child is HIV-positive!!!'

Fig. 5.8: Protection of feet

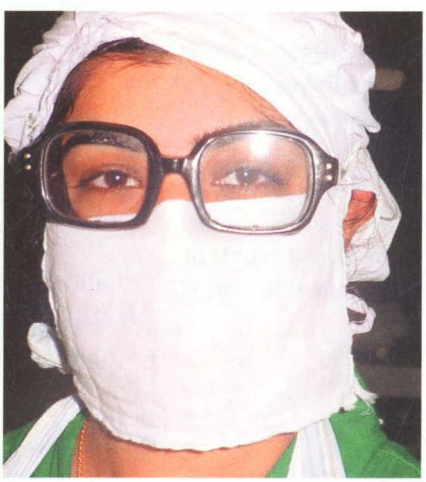


Fig. 5.9: Eye protection

Fig. 5.10: Double glove

3. HIV infection
4. Lymphoproliferative disease (Key Box 5.5)
5. **Kaposi's sarcoma** can affect skin surface, gastrointestinal tract from mouth to anus, lungs, etc. It presents as pigmented multifocal skin lesions.

IV. Oesophageal ulcers can present as dysphagia and odynophagia. Endoscopy should be done by using a glove and the scope should be washed thoroughly with soap and water and immersed in cetrimide solution for 15 minutes.

V. Lymphadenopathy: Very often surgeons are called for lymph node biopsy for evaluation of fever or generalised lymphadenopathy. All universal precautions have to be taken.

KEY BOX 5.5

HIV-ASSOCIATED LYMPHOMAS

1. Lymphomas in HIV patients—'B' symptoms and intra-abdominal and extranodal involvement including lung, bone marrow and liver are common.
2. HIV-associated lymphomas are ***Diffuse Large Cell*** (DLC) or Burkitt's lymphoma.
3. HIV-associated lymphomas present as ascites/pleural effusion, etc.
4. Treatment should be HAART (Highly Active Anti Retroviral Therapy) with standard chemotherapy and granulocyte-colony—stimulating factor.

Risk of HIV transmission

- Risk of HIV transmission from the patient to a surgeon is very low but it is dangerous. In US, by December 31, 2001, only 6 cases of surgeons with HIV seroconversion from a possible occupational exposure is documented by Centre for Disease Control and Prevention (CDC) (out of a total of 4,70,000 reported cases of HIV).
- Routine use of barriers (gloves and goggles), washing hands before and after examination of patient and careful handling of sharps and needles will greatly reduce transmission of the disease.
- Postexposure prophylaxis should begin immediately.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- Most of the diseases in this chapter are not of much interest to us other than HIV. I have given a new Key Box—**FACE MASK**. Detailed drug treatment of HIV is in medical books.
- An interesting clinical note on 'nerve abscess' has been added.

MULTIPLE CHOICE QUESTIONS

1. Which of the following statement is false in actinomycosis?
A. Sulphur granules are gram-positive mycelia surrounded by gram-positive clubs
B. *Actinomyces israelii* is an anaerobic organism
C. Organism has both bacterial and fungal characteristics
D. Multiple subcutaneous nodules over the jaw are characteristic

2. Following are true for treatment of actinomycosis *except*:
A. Crystalline penicillin is the drug of choice
B. Tetracyclines have no role to play
C. Surgery may be required for abdominal actinomycosis
D. Ceftriaxone can be used for pulmonary actinomycosis

3. Following are features of tuberculoid leprosy *except:*
A. Bacilli are few
B. Hypopigmented skin patch is seen
C. Nerve thickening is often a finding
D. Face and nose involvement is diagnostic of the condition

4. Hutchinson's triad does not include following:
A. Interstitial keratitis
B. 8th nerve deafness
C. Chancre
D. Narrow-edged wide spaced permanent incisors

5. Gummatous ulcer has following features *except:*
A. Commonly seen in the subcutaneous tissues
B. Floor has wash leather slough
C. Healing results in hypertrophic scar
D. It has punched out edges

6. Following are features of secondary syphilis *except:*
A. Vesicles all over the body
B. Coppery red rashes
C. Snail track ulcers
D. Epitrochlear node enlargement

7. Characteristic feature of primary genital chancre in syphilis is:
A. Seen after 48 hours of sexual exposure
B. Multiple ulcers
C. Indurated ulcer with large rubbery inguinal nodes
D. Indurated, single, painless ulcer

8. Following are features of *Treponema pallidum except*:
A. It is a spirochete
B. It is a gram-negative organism
C. Visible in dark field illumination
D. It does not have helical structure

9. Following is true for cytomegalovirus (CMV) infections *except*:
A. It causes abdominal pain and colitis
B. It is related to chickenpox—herpes virus
C. It can be transmitted by blood transfusion and organ transplant
D. Not transmitted to child during pregnancy

10. HIV associated lymphomas can be following *except*:
A. It is B cell lymphoma
B. They are diffuse large cell
C. They can be Burkitt's lymphoma
D. Hodgkin's lymphoma

ANSWERS

1 A 2 B 3 D 4 C 5 C 6 A 7 D 8 D 9 D 10 D

6

Differential Diagnosis of Leg Ulcer and Pressure Sore

- Clinical examination
- Management
- Wound dressings
- Traumatic ulcer
- Venous ulcer
- Trophic ulcer
- Tropical ulcer
- Post-thrombotic ulcer
- Rare ulcers
- Diabetic ulcer foot
- VAC
- Maggot therapy
- Pressure sore
- What is new?/Recent advances

Introduction

Leg ulcers are one of the important topics in surgery. They can occur in children, adults and the elderly. No age is spared, no sex is spared. Varying aetiological factors and presence of complicated systemic diseases make the treatment of ulcers very difficult. Chronic ulcers in old people definitely cause considerable morbidity and diabetic ulcer of the leg can cause life-threatening complications such as diabetic ketoacidosis and septicaemia. Hence, it is necessary to do a careful clinical examination of the ulcer to arrive at the diagnosis and plan for appropriate treatment.

Definition

An ulcer is a ***discontinuity of the skin or mucous membrane*** which occurs due to **microscopic death** of the tissues. Thus, ulcer can occur anywhere in the body (skin), oral cavity, penis (mucous membrane) or in the duodenum, intestine, etc. In this chapter, lower limb ulcers will be discussed.

Classification

Ulcers can be classified based on the **pathology** or **clinical features**.

I. Pathological classification (Key Box 6.1)

A. Nonspecific ulcers

1. **Traumatic:** Trauma can be mechanical. This is the most common cause of leg ulcer. It can be physical trauma due to burns or radiation. It can also be due to chemicals such as acids.
2. **Venous ulcers:** They include varicose ulcers and post-thrombotic ulcers which can occur following deep vein thrombosis.
3. **Arterial ulcers:** Following are a few examples of arterial ulcers:
   - **Buerger's** disease—common
   - **Atherosclerotic** vascular disease—common
   - **Vasospastic** disorders such as Raynaud's disease—uncommon
   - **Martorell's ulcers** or hypertensive ulcers—rare
   - Patients with rheumatoid arthritis can develop leg or foot ulcers due to vasculitis.
4. **Neurogenic ulcer** (neuropathic ulcer, trophic ulcers)
   - Leprosy and diabetes are the common causes

KEY BOX 6.1

PATHOLOGICAL CLASSIFICATION OF ULCER

A. Nonspecific ulcers
   1. Traumatic
   2. Venous
   3. Arterial
   4. Neurogenic—trophic
   5. Tropical
   6. Diabetic
   7. Blood dyscrasias

B. Specific ulcers

C. Malignant ulcers

- Paraplegia, meningomyelocoele, posterior tibial nerve injury, tabes dorsalis are the other causes.

5. **Tropical ulcer:** It is a rare ulcer due to malnutrition associated with infection caused by Vincent's organisms.
6. **Diabetic ulcer foot or diabetic ulcer leg**
7. **Blood dyscrasias:** Sickle cell anaemia, thalassaemia, leukaemia, etc. can produce recurrent ulcerations in the leg.

B. Specific ulcers

There are due to specific type of organisms, e.g. tubercular ulcers, syphilitic ulcers, actinomycotic ulcers.

C. Malignant ulcers

Malignant ulcers are squamous cell carcinoma, basal cell carcinoma, malignant melanoma. Malignant ulcers are discussed in Chapter 11.

II. Clinical classification (Table 6.1)

CLINICAL EXAMINATION OF AN ULCER

Inspection

1. **Location of the ulcer**
   - Arterial ulcer: Tip of the toes[1], dorsum of the foot.
   - Long saphenous varicosity with ulcer: Medial side of the leg.
   - Short saphenous varicosity with ulcer: Lateral side of the leg just above the lateral malleolus.
   - Perforating ulcers: Over the sole at pressure points.
   - Nonhealing ulcer: Over the shin and lateral malleolus.
2. **Floor of the ulcer:** This is the part of the ulcer which is exposed or seen.
   - Red granulation tissue: Healing ulcer (Fig. 6.1)
   - Necrotic tissue, slough: Spreading ulcer (Fig. 6.2)
   - Pale, scanty granulation tissue: Tuberculous[2] ulcer.
   - Wash-leather slough: Gummatous ulcer
   - Part of the bone: Neuropathic ulcer
   - Nodular: Epithelioma
   - Black tissue: Malignant melanoma

Fig. 6.1: Traumatic ulcer with red granulation tissue in the floor—typical healing ulcer

Fig. 6.2: Slough—dead soft tissue—typical spreading ulcer in a diabetic patient

3. **Discharge from the ulcer**
   - Serous discharge: Healing ulcer
   - Purulent discharge: Spreading ulcer
   - Bloody discharge: Malignant ulcer
   - Discharge with bony spicules: Osteomyelitis
   - Greenish discharge: Pseudomonas infection
4. **Edge:** This is between the floor of the ulcer and the margin. ***The margin is the junction between the normal epithelium and the ulcer.*** It represents the **areas of maximum cellular**

Table 6.1 Clinical classification

A. Spreading	B. Callus	C. Healing
No granulation tissue	Pale granulation tissue	Red granulation tissue
Plenty of discharge	Serous discharge	Minimal serous discharge
Excessive slough	Slough present	Slough absent
Surrounding area inflamed and oedematous	Induration at the base, edge and surrounding area	Signs of inflammation are minimal
Purulent smell present	Smell can be present	Smell is absent

[1]When there is a block in the pipelines supplying water, distal houses suffer the maximum. Is it not?
[2]It is described as **apple jelly granulation tissue**.

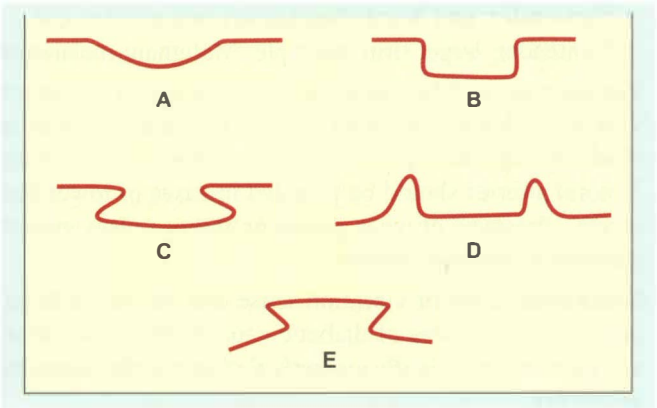


Fig. 6.3: Different types of edges of ulcer

activity. If destruction dominates as in spreading ulcers, the edge is inflamed, oedematous and angry-looking (*stage of extension*). When ulcer shows evidence of healing, the edge will be bluish due to granulation tissue covered with thin epithelium (*stage of transition*). In a healed ulcer, the outermost part of the edge is whitish due to fibrosis (*stage of repair*)—for types of edge (Fig. 6.3).

- **Sloping edge** is seen in all **healing ulcers** such as traumatic ulcers, venous ulcers (Fig. 6.4).
- **Punched out edge** is seen in **gummatous ulcers** and **trophic ulcers**. Gummatous ulcers have punched out edge due to endarteritis obliterans caused by syphilitic organisms. Chronic nonhealing ulcers may also have punched out edges (Fig. 6.5).
- **Undermined edge** is seen in **tuberculous** ulcers, probably due to more destruction of subcutaneous tissues than the skin. The edge is classically thin and bluish in colour (Fig. 6.6).
- **Raised edge** (beaded edge) is seen in rodent ulcers or **basal cell carcinoma** (Fig. 6.7).
- **Everted edge** (rolled out) is diagnostic of **squamous cell carcinoma**. The edge grows very rapidly and it occupies the normal skin and thus gets averted (Fig. 6.8).

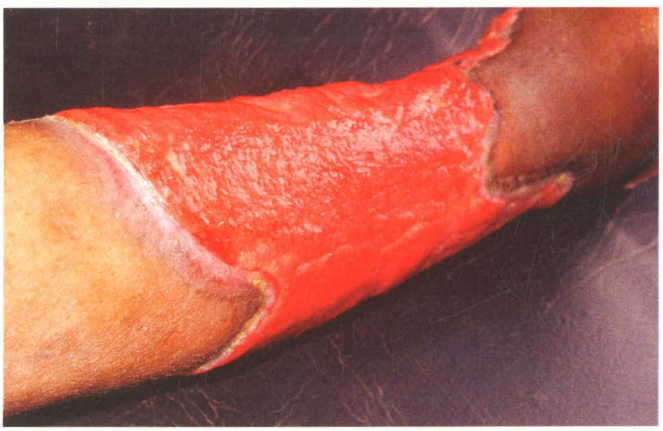


Fig. 6.4: Sloping edge

Fig. 6.5: Punched out edge—neuropathic ulcer (classically described for gummatous ulcer)

Fig. 6.6: Undermined edge—tubercular sinus/ulcer

Fig. 6.7: Elevated edge—basal cell carcinoma (*Courtesy:* Prof Vidyadhar Kinhal, HOD, Surgery, VIMS, Bellary, Karnataka)

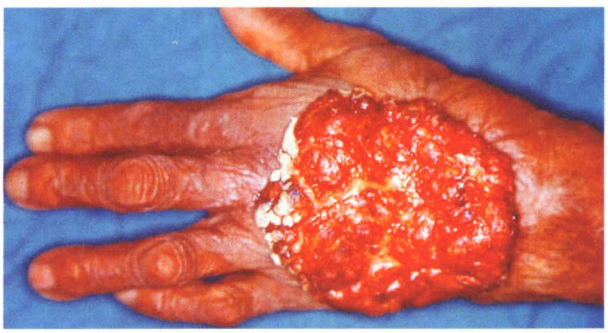


Fig. 6.8: Everted edge—squamous cell carcinoma

5. Surrounding area
- Thick and pigmented: Varicose ulcer.
- Thin and dark: Arterial ulcer.
- Red and oedematous: Spreading ulcers like diabetic ulcer.
- Scar around the ulcer: Marjolin's ulcer.

Palpation

1. **Edge:** Induration (hardness) of the edge is characteristic of squamous cell carcinoma. Some degree of induration can also be seen in chronic ulcers and long-standing varicose ulcers. Induration occurs due to extensive fibrosis. It is said to be a host defense mechanism. *By causing fibrosis, lymphatic spread is delayed.* Tenderness of the edge is characteristic of infected ulcers and arterial ulcers.
2. **Base:** *It is the area on which ulcer rests*. Pick up the ulcer between thumb and index finger and tissues beneath can be appreciated. If the ulcer cannot be lifted up, the base cannot be made out. The base can be tendons, muscles or bone depending upon the site of ulcer. ***Marked induration at the base is diagnostic of squamous cell carcinoma.*** Hunterian chancre is a benign ulcer and produces significant induration. Hence, it is also called a hard chancre (Key Box 6.2).
3. **Mobility:** Gentle attempt is made to move the ulcer to know its fixity to the underlying tissues. Malignant ulcers are usually fixed, whereas benign ulcers are not.
4. **Bleeding:** Malignant ulcer is friable like a cauliflower. On gentle palpation, it bleeds. **Granulation tissue** as in a healing ulcer also bleeds[1].
5. **Surrounding area:** Thickening and induration is found in squamous cell carcinoma. Tenderness and pitting on pressure indicates spreading inflammation surrounding the ulcer.

Relevant clinical examination

1. Regional lymph nodes
- Tender and enlarged: Acute secondary infection
- Nontender and enlarged: Chronic infection
- **Nontender and hard: Squamous cell carcinoma**
- Nontender, large, firm, multiple: Malignant melanoma

KEY BOX 6.2

INDURATION
- It means hardness
- Maximum induration—Squamous cell carcinoma
- Minimal induration—Malignant melanoma
- Brawny induration—Abscess
- Cyanotic induration—Chronic venous congestion as in varicose ulcer
- The base and the surrounding area should be examined for induration.

2. **Peripheral vessels:** Detailed examination of peripheral vessels is discussed under peripheral vascular disease. However, dorsalis pedis, posterior tibial, popliteal and femoral arteries should be palpated in cases of lower limb ulcers. Presence of weak pulses or absent pulses indicate peripheral vascular disease.
3. **Sensations:** Loss of vibration sense and loss of ankle jerk occurs early in cases of diabetic neuropathy. Later, touch and pain are lost. ***Totally anaesthetic feet are characteristic of leprosy***.
4. **Function of the joint:** Movements of the involved joint are restricted either due to pain, involvement of the joint or due to infiltration into the joint by malignant ulcers.
5. **Varicose veins:** If present, it is most probably a varicose ulcer. However, A-V fistula can present as distal ulcers with arterialisation of veins and a continuous murmur.

Systemic examination

1. **Central nervous system** (CNS) and spine in neuropathic ulcers. There may be gibbus as in cases of TB spine or operated scar due to myelomeningocoele, etc. *See* clinical notes.

CLINICAL NOTES

An 18-year-old girl with a nonhealing trophic ulcer was examined by a postgraduate student. He gave a diagnosis of trophic ulcer (neuropathic ulcer) due to leprosy as first diagnosis followed by polyneuropathy. He failed. It was a case of myelomeningocoele and the candidate had not examined the spine! The patient had an operated myelomeningocoele.

2. **Splenomegaly** in blood dyscrasias such as in early stages of sickle cell anaemia.
3. **Cardiovascular system** (CVS) may reveal murmur as in cases of arteriovenous fistula or features suggestive of cardiac diseases (for summary *see* Key Box 6.3).

Investigations

1. **Complete blood picture:** Hb%, TC, DC, ESR, peripheral smear
   - Low Hb% is found in chronic ulcer. It is either nutritional or due to frequent blood loss during dressings as in diabetic ulcer.
   - High total count indicates infection.
   - Peripheral smear is done to rule out anaemia and sickle cell disease.

[1]Granulation tissue is made up of capillaries and fibroblasts. Hence, it gives rise to fresh blood loss.

KEY BOX 6.3

CLINICAL EXAMINATION OF AN ULCER

Inspection

- Location, size, shape, floor, edge, discharge, surrounding area.

Palpation

- Tenderness, local rise of temperature, bleeding on touch, consistency of the ulcer, edge, surrounding area—oedema, mobility.
- Regional lymph nodes
- Sensations
- Pulsations
- Function of the joint
- Systemic examination

2. **Urine** and blood examination to rule out diabetes
3. **Chest X-ray:** PA view to rule out pulmonary tuberculosis
4. **Pus** for culture/sensitivity
5. **Doppler/duplex scan/lower limb angiography** in cases of arterial diseases
6. **X-ray of the part** to look for
   - Osteomyelitis—common in diabetic ulcers
   - Periostitis tibia—common in varicose ulcers
7. **Biopsy:** Nonhealing/malignant ulcers

Treatment of the ulcers

It can be discussed under the following headings

1. Treatment of spreading ulcers
2. Treatment of healing ulcers
3. Treatment of chronic ulcers
4. Treatment of the underlying disease

1. Treatment of spreading ulcers

After obtaining pus culture/sensitivity report, appropriate antibiotics are given. Many solutions are available to treat the slough, such as hydrogen peroxide and **Eusol**[1].

- Hydrogen peroxide (diluted) when poured over the wound, liberates nascent oxygen which bubbles out and helps in separating the slough. Eusol also separates the slough. ***There are reports that H_2O_2 and Eusol can cause more damage. Hence, they are no longer used.***
- Partially separated slough needs to be removed daily or on alternate days, in the wards.
- **Excessive granulation tissue** or pouting granulation tissue (proud flesh) needs to be decapitated by excision or by application of copper sulphate or silver nitrate solution.

  Thus, by repeated dressings, slough gets separated and discharge becomes minimal, resulting in a healing ulcer with healthy red granulation tissue. Management thereafter is like that of a healing ulcer.
- *See* Table 6.2 for various ulcer dressing.

[1]Eusol—Edinburgh University Solution (hypochlorite solution).

2. Treatment of healing ulcers

- Regular dressings are done for a few days with antiseptic creams such as liquid iodine, zinc oxide or silver sulphadiazine preparation.
- ***A swab is taken to rule out the presence of Streptococcus haemolyticus which is a contraindication for skin grafting.***
- If the ulcer is small, it heals by itself with epithelialisation from the cut edge of ulcer.
- If the ulcer is large, free split skin graft is applied as early as possible (Key Box 6.4).

KEY BOX 6.4

ADVANTAGES OF SPLIT SKIN GRAFT

- Wound healing occurs fast
- Secondary infection is avoided because of early skin cover
- It prevents contractures
- It prevents Marjolin's ulcer—squamous cell carcinoma arising from scar tissues

3. Treatment of chronic ulcers (Table 6.2)

These are the ulcers which do not respond to conventional methods of treatment. Some special forms of treatment are available but their usefulness is doubtful. They are as follows:

- Infrared radiation, short-wave therapy, ultraviolet rays decrease the size of the ulcer.
- Amnion helps in epithelialisation.
- Chorion helps in formation of granulation tissue. These ulcers ultimately may require skin grafting (Fig. 6.9).

Fig. 6.9: After epidermal growth factor for 4 days, wound became very vascular

4. Treatment of the underlying disease (*vide infra*)

Differential diagnosis (Table 6.3)

TRAUMATIC ULCER

They can occur anywhere in the body. However, they are more common where skin is closely applied to bony prominences, e.g. shin, malleoli, over which there are no muscles. They are usually single, very painful ulcers of healing type (Fig. 6.10).

Table 6.2 Classes of wound dressings, debridement agents and skin replacements currently available

Class	Composition	Characters/Functions	Commercial examples/ comments
1. Debriding agents			
• Hydrogen peroxide	H_2O_2 releases nascent oxygen which bubbles out and slough comes to surface.	Destroys anaerobic bacteria; heat generated causes vasoconstriction and haemostasis, frothing brings debris to the surface	Both these are not favoured today (Eusol is Edinburgh University Solution)
• Eusol	Hypochlorite solution	Mild debriding action	
2. Polymeric films	Plastic (polyurethane); semipermeable	Allows water vapour permeation; adhesive	Opsite; Tegaderm
3. Hydrocolloid dressings	Hydrophilic colloidal particles and adhesive. It is impermeable to fluids and bacteria	Absorbs fluid; necrotic tissue autolysis; little adherence; occlusive forms complex structures with water and aids in atraumatic removal of the dressing, hydrocol allows a high rate of evaporation without compromising wound hydration	Duoderm, Intrasite
4. Alginates	Polymer gel contains mannuronic acid and glucuronic acid—Seaweed polymer that forms a gel when it absorbs fluid	Absorbs exudates; nonadherent, nonirritating, requires a cover dressing (permeable) Along with silver[1], antimicrobial action against MRSA and pseudomonas.	Algisorb, Sorbsan should not be used in presence of hepatic or renal impairment
5. Miscellaneous			
• Gauze	Woven cotton fibres	Permeable with desiccation; debridement; painful removal	
• Tulles			
6. Medicated dressings	Medications	Increased epithelialisation by 25–30%	Zinc oxide, Neomycin, Bacitracin, Zinc
• Impregnated gauzes	Fine mesh fabric (silicone, nylon) with dermal porcine collagens)	Nonadherent; semipermeable	Biobrane II
7. Platelet-derived growth factor (PDGF)	Acts through tyrosine kinase receptor	Stimulates growth of cells and angiogenesis; increases granulation tissue; stimulation of repair—used when blood supply is good, classically neuropathic nonhealing ulcers of diabetics	Plemin, Regen-D
8. Endothelial-derived growth factor (EDGF)	Acts through tyrosine kinase receptor	Stimulates growth of cells and angiogenesis; increases granulation tissue; stimulation of repair—used when blood supply is good, classically neuropathic nonhealing ulcers of diabetics	Costly

Fig. 6.10: Traumatic multiple ulcers over the shin—classical site

With proper dressings and antibiotics, they usually heal within 5–7 days.

- **Footballer's ulcer** is the name given to those nonhealing ulcers which occur in the leg over the shin due to direct trauma caused by the football. Sometimes, these ulcers may take a long time to heal. If not treated properly, it gets adhered to the bone.

VENOUS ULCER (Fig. 6.11)

- It occurs due to increased venous hydrostatic pressure.
- Located on the medial side of lower one-third of the leg in cases of long saphenous varicosity and on the lateral aspect of the leg in short saphenous varicosity.
- It is shallow and superficial.
- Never penetrates deep fascia
- Usually painless unless it is infected or causes periostitis tibia

[1]Silver is antimicrobial and calcium is haemostatic.

Table 6.3 Causes of ulcer

Common causes	Uncommon causes	Rare causes
Traumatic ulcer	Neurogenic ulcer	Martorell's ulcer
Varicose ulcer	Tropical ulcer	Bazin's disease
Arterial ulcer	Post-thrombotic ulcer	
Diabetic ulcer	Malignant skin ulcer	

- Shows evidence of healing
- Usually associated with varicose veins
- Typically lower leg around the ulcer is pigmented.

ARTERIAL ULCER (ISCHAEMIC ULCER)

They are very painful and occur in young patients who have Buerger's disease or occur in elderly patients due to atherosclerotic vascular disease. It commonly occurs on the tips of toes and fingers (Figs 6.12 and 6.13). The ulcer is dry, deep and penetrates deep fascia. Evidence of chronic ischaemia in the rest of the foot clinches the diagnosis (Table 6.4 for differences between arterial and venous ulcers).

Fig. 6.11: Varicose ulcer-pigmentation is characteristic

Fig. 6.12: Ischaemic ulcer on the dorsum of the foot—typical site

Fig. 6.13: Polycythaemia causing gangrene of toes

NEUROGENIC ULCER, NEUROPATHIC ULCER, TROPHIC ULCER (Figs 6.14 and 6.15)

This type of ulcer develops in an anaesthetic limb. The causes of neuropathy are:

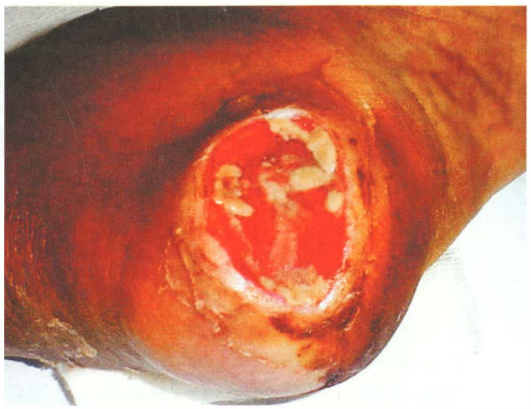
- Diabetic neuropathy
- Meningomyelocoele
- Leprosy
- Alcoholic neuropathy
- Nerve injuries
- Transverse myelitis
- Ulcer develops on the pressure points such as beneath heel, beneath the first and fifth metatarsals and gluteal region (decubitus ulcer). It develops as a callosity, gets infected, suppurates and leaves a central hole discharging pus. Slowly, it burrows deep inside, may involve bone and cause osteomyelitis. Hence, it is also called **perforating ulcer**.
- Trophic ulcers are caused by inadequate blood supply, malnutrition and neurological deficit. They are also included in this group.

Treatment

- **Immobilisation of the foot in a plaster of Paris** posterior slab with a walking boot almost cures the ulcer within 2–3 weeks, provided the primary disease is also controlled, e.g. leprosy. If the ulcer is nonhealing with slough, initial management should include desloughing agents and surgical removal of the slough.

Table 6.4 Differences between arterial and venous ulcers

	Arterial ulcer	Venous ulcer
Location	Tips of toes	Medial or lateral side of leg
Pain	Very painful	Absent
Number and shape	Many and irregular	Single and oval
Depth	Deep, penetrates deep fascia	Superficial, does not penetrate deep fascia
Pigmentation	Not a feature	Usually present
Nature of the vessels	Peripheral pulses are weak or absent; veins are not dilated	Peripheral pulses are normal; veins are dilated



Figs 6.14 and 6.15: Neuropathic ulcer—classical site over the heel

TROPICAL ULCER

It occurs in tropical countries. The precipitating factors are:

- Malnutrition
- Humid zones
- Poor immunity
- Trauma or insect bite

The infection is caused by Vincent's organisms like bacteroides, *B. fusiformis* and *Borrelia vincentii*. It starts as a pustule with extensive inflammation. The pustule bursts and the ulcer spreads rapidly and causes destruction of surrounding tissue. Hence, it is also called **phagedenic ulcer**[1]. The edges are undermined, floor contains slough, and there is copious seropurulent discharge. Healing is delayed for days to a month. Metronidazole may be quite useful in bringing down the inflammation. Broad-spectrum antibiotics may also be required in cases of secondary infections. If healing takes place, it leaves behind a scar.

POST-THROMBOTIC ULCER

It occurs due to deep vein thrombosis. It may affect calf veins or it may be due to femoral vein thrombosis. It is an example of venous ulcer or gravitational ulcer.

Precipitating factors

- Accidents involving lower leg
- Following childbirth
- After abdominal operation.

Clinical features

- Bursting pain in the limb
- Extensive induration of the leg or thigh depending upon site of thrombosis
- The ulcer is nonhealing with scanty granulation tissue.
- The ulcer is deep and always infiltrates deep fascia.
- Due to increased hydrostatic venous pressure, the part is significantly indurated (cyanotic induration), pigmented and thickened with a rise in local temperature.
- The ulcer is not associated with superficial varicosity.
- **Homan's sign:** It is positive in calf vein thrombosis. It is elicited by forcible dorsiflexion of the foot with the knee extended causing pain in the region of calf.
- **Moses's sign:** Squeezing of the calf muscles from side to side also produces pain.

  These two signs are positive in acute cases.

Treatment

- Rest and elevation of the leg
- Appropriate antibiotics
- Elastic crepe bandage

With conservative treatment for a few days to a few weeks, veins may get recanalised and the ulcer may heal. The treatment can be very difficult (*see* Chapter 10 on varicose veins).

RARE ULCERS[2]

MARTORELL'S ULCER

- Affects elderly patients over the age of 50 years.
- Commonly affects hypertensive patients. Hence, the name **hypertensive ulcers**.
- Atherosclerosis is also a precipitating factor even though all peripheral pulses are usually present.
- It occurs due to sudden **obliteration of end-arterioles** of the skin on the back or outer side of calf region.
- Severe pain, ischaemic patch of skin which later develops into a deep punched out nonhealing ulcer is a clinical feature.
- Healing is delayed due to vascular insufficiency.

[1]Phagedenic (to eat). Rapidly spreading, ulcerative, destructive lesion. It can occur in the oral cavity and also over the penis.

[2]Students should not offer these ulcers as clinical diagnosis. They are rare ulcers, with rare clinical interest.

BAZIN'S ULCER

- These ulcers exclusively occur in **young females** and occur in the lower third of leg and ankle region.
- Usually seen in those patients who are **obese** with thick ankles and abnormal amount of subcutaneous fat.
- It begins with erythematous purplish nodules (hence the name **erythrocyanosis frigida**) on the calves which later rupture producing nonhealing ulcer.
- Aetiology of these ulcers is not clear. It is supposed to be due to **ischaemia of lower leg** due to spasm of branches of posterior tibial and peroneal arteries. These vessels are **abnormally sensitive to hot and cold weather** similar to Raynaud's disease. In some cases, tubercle bacilli have been isolated, with ulcers responding to antituberculous treatment.
- These ulcers are managed conservatively.
- **Sympathectomy** may be beneficial in those patients who are hypersensitive to weather changes.

DIABETIC ULCER FOOT

- Risk factors are given in Key Box 6.5

Diabetic patients are more prone for development of ulcers in the foot because of the following reasons:

1. **Neuropathy[1]:** It commonly manifests after about 10 years of diabetes. Neuropathy can be distal and diffuse with a stocking type of distribution. ***Nerve damage is due to formation of sorbitol from sugar.*** Sorbitol causes demyelination of large fibres. New **arteriovenous communications** open beneath skin, diverting nutrient flow away from it. This ischaemic tissue is vulnerable for infection causing nerve ischaemia. Loss of vibration sense and deep tendon reflexes occur early. Later, joint position, touch, pain and temperature sensations are lost. As a result of this, trophic ulcer develops. It progresses and can penetrate deeper and deeper. Very often the patient is unaware of this.
   - Diabetic neuropathy of the tibial nerve is dangerous. ***Clawing of the toes and hammer toe result due to paralysis of the intrinsic muscles of the foot.*** Sensation is absent over the entire sole of the foot due to involvement of medial and lateral plantar nerves. These two factors predispose to the pressure sore over the plantar surface of the head of the metatarsals.
   - **Charcot's arthropathy (joints)** is a form of arthritis seen in neuropathic foot and leads to severe deformity.
   - **Autonomic neuropathy** leads to absence of sweating and gives rise to anhydrotic skin (Key Box 6.6).

[1]Burning or tingling in plantar aspect of forefoot radiating to toes—**Morton's interdigital neuroma**.

KEY BOX 6.5

RISK FACTORS FOR DIABETIC ULCER FOOT

- Male above 50 years
- DM of more than 10 years duration
- Blood glucose levels not controlled
- Peripheral neuropathy
- Abnormal structure of foot
- Peripheral vascular disease
- Smoking and hypertension
- Increased level of lipids
- Genetic factors

KEY BOX 6.6

DIABETIC NEUROPATHY

- Loss of vibration and touch
- Loss of pain and temperature
- Dead like feeling in the feet
- Sensation of walking on the 'sand'
- Susceptible for repeated trauma
- Nylon monofilament test is routinely used to test neuropathy by pressing in on the foot/sole

PEARLS OF WISDOM

Autosympathectomy is a feature of diabetic foot ulcers.

- Various factors of neuropathy contribute to changes in configuration of foot and change in gait. Foot becomes shorter and wider. Longitudinal arch becomes flat. New pressure points develop resulting in ulcers in the foot (Key Boxes 6.6 and 6.7).

2. **Resistance to infection** is lowered in diabetes mellitus due to altered immune system. Uncontrolled diabetic patients are more susceptible for infection. Even though leukocytosis occurs in diabetic patients with infection, the **phagocytic activity of the leukocytes is greatly reduced**.
   - **In ketoacidosis, granulocyte mobilisation is impaired** and chemotaxis is also reduced. Thus, they are susceptible for polymicrobial and fungal infection.

KEY BOX 6.7

EFFECTS OF DIABETIC NEUROPATHY

- Loss of sensation
- Loss of sweating
- Loss of muscle strength
- Loss of curvature of foot
- Loss of normal joint position
- Loss of elasticity of skin

3. **Atherosclerosis:** Diabetic angiopathy involving major vessels results in ischaemia of the foot **(macroangiopathy)**. They have accelerated atherosclerosis. In addition, it also produces small vessel disease in the form of nonspecific thickening of the basement membrane. It is described as **microangiopathy** (Key Box 6.8).

Thus, neuropathy or microangiopathy singly or in combination with secondary infection favours the development of diabetic ulcer. Ulcer starts due to minor trauma such as thorn prick, trimming of the nail or due to shoe bite. It may also start as a callosity in the sole of the neuropathic foot. Hence, utmost care should be taken to protect foot.

KEY BOX 6.8

DIABETES AND ATHEROSCLEROSIS

- 16% of diabetic patients have PVD (peripheral vascular diseases)
- Collateral circulation is reduced in diabetics
- Atherosclerotic plaque causes increased ulceration and cracking
- More severe the diabetes, more severe will be **peroneal occlusive disease**

Sequence of events in diabetic ulcer foot—Complications (Fig. 6.16)

1. Following an injury or due to infection, an ulcer develops along with swelling and oedema of the leg—**Stage of cellulitis**.
2. Cellulitis stage takes up a virulent course, spreads deeper and also upwards along fascial planes—**Stage of spreading cellulitis**.
3. Secondary infection caused by mixed organisms along with anaerobes and nonclostridial gas-forming organisms produce multiple abscesses—**Stage of abscesses**.

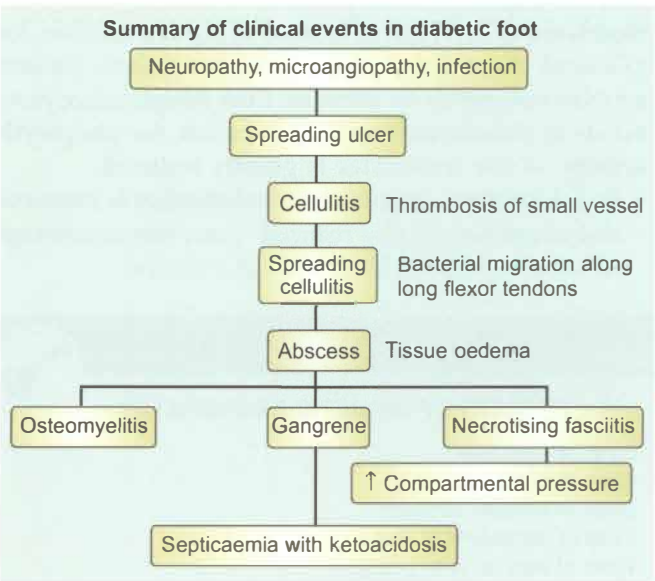


Fig. 6.16: Various stages of infection of the leg in diabetic patients

4. Tense oedema along with vascular compromise which is already existing produces ischaemia and gangrenous patches of skin, toes, etc.—**Stage of gangrene**.
5. Infection involves deeper tissues such as bone, producing osteomyelitis—**Stage of osteomyelitis**.
6. Untreated cases develop rapidly spreading cellulitis and **gangrene of the limb** producing septicaemia and diabetic ketoacidosis—**Stage of septicaemia**.

Investigations

1. Complete blood picture usually demonstrates high total count with low Hb% (infection).
2. Blood and urine sugar estimations
3. Pus for culture/sensitivity
4. X-ray of the foot to rule out osteomyelitis which may be the cause for chronicity of the ulcer (Fig. 6.17).
5. Liver function test (LFT), ECG, chest X-ray, blood urea, serum creatinine as a routine in diabetic patients (remember diabetes is a systemic disease).
6. Lower limb arterial duplex scan is an important investigation to check the patency of vessels.
   - Turbulent blood flow (normal is laminar flow wider spectrum frequency and loss of diastolic forward flow is typically seen in diabetic foot ulcer.
7. Hand held Doppler and measurement of ankle brachial index (ABI) is a bedside test.
8. Aortogram when you decide reconstruction (CT angiogram).

Fig 6.17: Destruction of metatarsal bone and dislocation of metatarsal joint

Grading of diabetic ulcer foot (Key Box 6.9)

KEY BOX 6.9

MODIFIED WAGNER GRADING SYSTEM OF DIABETIC ULCER FOOT

Grade 0: No skin changes
Grade 1: Superficial ulcer
Grade 2: Ulcer extension
  a. Involves ligament, tendon, joint capsule or fascia
  b. No abscess, no osteomyelitis
Grade 3: Deep ulcer with abscess or osteomyelitis
Grade 4: Gangrene of the portion of forefoot
Grade 5: Extensive gangrene of foot

Treatment[1] of diabetic ulcer of the foot

It can be discussed under five headings:

- Control of diabetes
- Control of infection
- Local treatment of the ulcer
- Various types of surgery for diabetic ulcer of the foot
- Care[2] of the patient as a whole

1. **Control of diabetes:** It is an important part of the treatment of diabetic ulcer of the foot. Diabetes precipitates infection which worsens the diabetic status. These ulcers are better managed, at least in the initial period using insulin rather than oral antidiabetic drugs. Inj. plain insulin is given 3–4 times/day depending upon the requirement. With availability of **glucometer** and estimation of **GRBS**, sliding scale method based on urine sugar estimation has become obsolete.
2. **Control of infection:** Once culture/sensitivity report is available, appropriate antibiotics are started. Commonly gram-positive, gram-negative and anaerobic infection exist. Triple antibiotics may have to be continued for a long time depending upon the nature, type and severity of infection. ***Presence of high grade fever with chills and rigors suggests development of multiple abscess pockets that need to be drained rather than indiscriminate change and usage of antibiotics.*** If infection is not controlled properly, ketoacidosis can occur.
3. **Local treatment of diabetic ulcer foot:** Diabetic ulcer is a nonhealing ulcer. Hence, initial treatment is debridement/dressings or iodine solution and when the ulcer is converted into a healing ulcer, with pink granulation tissue, a split skin graft is applied. Small ulcers heal by granulation tissue (Fig. 6.18). Dakin's solution is a chlorine releasing agent. It is bactericidal and it loosens necrotic tissue. Collistin solution contains 75 mg colistin in 50 ml normal saline, can be used if Pseudomonas and anaerobic organism are present.
4. **Various types of surgery[3] for diabetic foot** (Table 6.5).

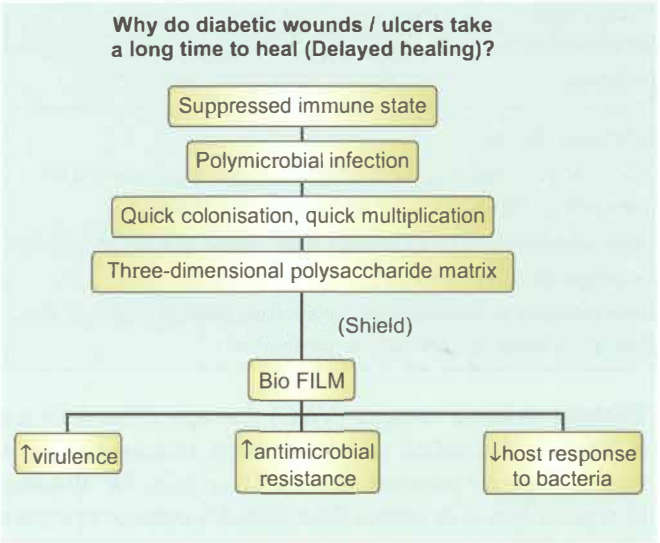


Fig. 6.18: Delayed healing of diabetic ulcers

5. **Care of the patient as a whole:** Recovery and healing of diabetic ulcer of the foot may range from a few weeks to a few months. During this period there are various other aspects to be looked after apart from infection and insulin (Table 6.6).
6. **Revascularisation of foot in diabetic patients**
   - Ankle brachial pressure index is ***misleading*** because of calcified vessels (incompressible).
   - **Duplex ultrasound** assesses both anatomical and functional abnormality. If there is stenosis, peak systolic velocity ratio will be greater than 2 across arterial lesion.
   - **Angiography** is still the gold standard.
   - For short stenotic lesions, **balloon angioplasty** with or without stent placement is the treatment.
   - **Infrainguinal bypass surgery**—popliteal to tibial or pedal artery using long saphenous vein.
   - Even after successful surgery, amputation rate is about 35%. 2 year patency rate is around 70%.

Table 6.5 Aim—to save the leg

Spreading ulcer with slough	Debridement
Healing ulcer	Skin grafting/flaps (reverse sural flap)
	Incision and drainage
Abscess	Disarticulate toe
Gangrene toe	Excision of metatarsal bones
Involvement of metatarsal bones	Forefoot amputation
Gangrene confined to toes	Multiple fasciotomy/decompression/debridement
Spreading cellulitis/necrotising fasciitis	Amputation below knee or above knee.
Spreading cellulitis with gangrene	Revascularisation, infrainguinal bypass
Ischaemic limb	

[1]Even a small diabetic ulcer should be treated properly. Otherwise a patient may have to 'pay through his foot' for it.
[2]This is an important aspect often forgotten by the treating physicians and surgeons.
[3]Unfortunately in some patients, in an attempt to save the limb, all these surgeries will be done and at last, they may end up with amputation of the leg.

Table 6.6 Various problems and their solutions

Problems	Solutions
Nutritional factors	Diabetic diet should be given
A bedridden patient may have difficulty in passing urine, situation worsened by pre-existing benign prostatic hypertrophy	Catheterisation (Foley's catheter) under aseptic measures, frequent change of catheter and catheter care
Chest infection such as pulmonary tuberculosis or static pneumonia	Control of tuberculosis, pneumonia, chest physiotherapy
Development of bedsores	Frequent change of position and nursing care
More sensitive to carbohydrate metabolism, protein depletion and changes in water and electrolyte metabolism	Frequent checking of Hb%, total proteins, blood glucose and electrolyte levels, etc.

7. Vaccum assisted closure (VAC) therapy (Figs 6.19 and 6.20): It is also called vacuum therapy, *vacuum sealing* or topical negative pressure therapy. It is used for drainage to remove blood or serous fluid from a wound or operation site.

Technique

- Foam with an open-cell structure is introduced into the wound and a drain is kept and brought out by lateral multiple holes. The entire area is then covered with a transparent adhesive membrane, which is firmly secured to the normal healthy skin around the wound margin. The drain tube is connected to a vacuum, fluid is drawn from the wound through the foam into a bag (Fig. 6.19).
- The plastic membrane prevents the inflow of air and bacteria. It also allows a partial vacuum to form within the wound, facilitates the removal of fluid.

How does it work?

- **Negative pressure** assists with removal of interstitial fluid, decreasing localised oedema and increasing blood flow.
- This in turn decreases tissue bacterial levels.
- Mechanical deformation of cells occur and it results in protein and matrix molecule synthesis, which increases the rate of cell proliferation.

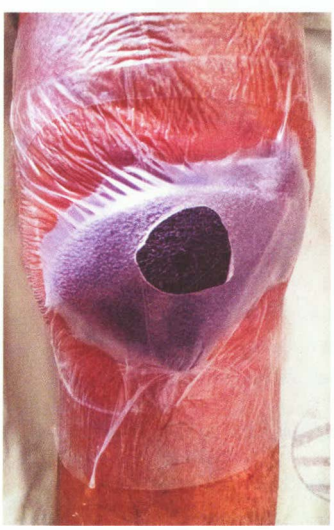


Fig 6.19: VAC

- **Equipment:** Microprocessor-controlled vacuum unit that is capable of providing controlled levels of continuous or intermittent sub-atmospheric pressure ranging from 25 to 200 mmHg.

Indications

- Venous ulcers, diabetic ulcers, large ulcers following necrotising fasciitis and any chronic ulcers.
- Patients with open fractures.
- Soft tissue injuries including sacral pressure ulcers, acute traumatic soft tissue defects and infected soft tissue defects following rigid stabilisation of lower limb fractures.
- Treatment of burns and wounds in the perineum, hand or axilla.

Contraindications

- Osteomyelitis—First treat osteomyelitis and then apply VAC
- Internal fistula
- Necrotic tissue in eschar
- Malignancy

8. Limited access dressing (LAD): A simple plastic sheet covers the wound. Intermittent suction is applied. It is cheaper when compared to VAC (Fig. 6.20).

Fig. 6.20: Limited access dressing

See Figs. 6.21 to 6.31 for various aspects of diabetic ulcer foot.

VARIOUS ASPECTS OF DIABETIC ULCER FOOT (Figs 6.21 to 6.31)

Fig. 6.21: Cellulitis leg

Fig. 6.22: Clawing of toes

Fig. 6.23: Wet gangrene with abscess foot

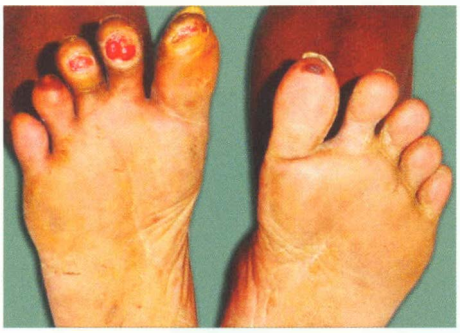


Fig. 6.24: This patient had heat burns following barefoot walking resulting in ulcers

Fig. 6.25 Pale granulation tissue and purulent discharge

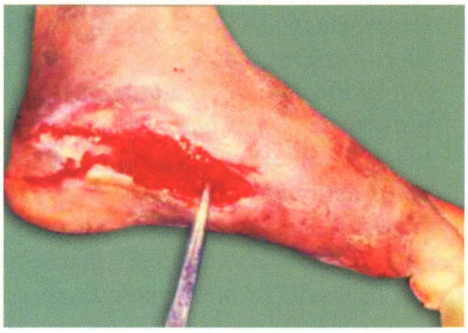


Fig. 6.26: Red granulation tissue-healing after control of diabetes and regular dressings

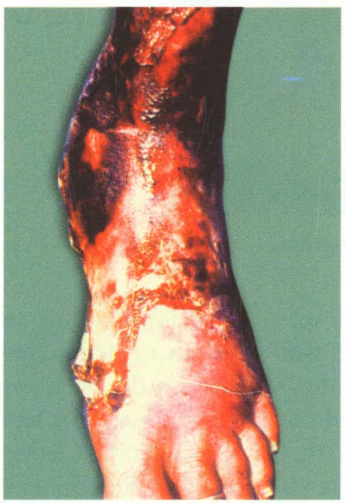


Fig. 6.27: Diabetic ulcer with moist gangrene. Patient had ketoacidosis with septicaemia. In spite of amputation, patient died

Fig. 6.28: Extensive diabetic ulcer of the foot—needed below knee amputation

Fig. 6.31: Below knee amputation (*see* Chapter 53 on amputation under operative surgery for more details)

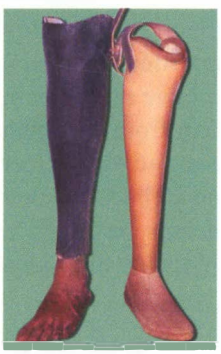


Fig. 6.29: Artificial limb prosthesis

Fig. 6.30: Microcellular rubber (MCR) chappals specially designed for patients with neuropathic foot. They avoid pressure points

Patient/public education to protect the foot (Key Box 6.10)

- Never walk barefoot, preferably use ***microcellular rubber shoes*** which are not only soft, but also allow oxygenation.
- Keep the foot dry after proper cleaning of the foot.
- Paring of the nails and trimming should be done carefully. If infection sets in, consult the physician at the earliest.
- **Avoid applying herbal/local medicines** or lotions to a corn. Consult the surgeons for treatment of the corn.
- Proper and regular control of diabetes by diet and frequent measurement of blood sugar.
- Do not consult the neighbours[1].
- Reassurance and good rapport with the treating doctors.
- Since these patients have peripheral neuropathy, they will **not be able to appreciate temperature of water**. Pouring hot water over the foot can cause burns without the patient being aware of it. Hence, a bystander has to check the temperature of water before being used by the patient.

Thus, the best way to avoid complications related to diabetes is by controlling the sugar levels with diet alone or diet with insulin or oral hypoglycaemic agents.

KEY BOX 6.10

PUBLIC EDUCATION IN DIABETIC PATIENTS

Barefoot walking should be avoided
Avoid herbal/local ointment application
Regular and **R**igorous control of diabetes with diet
Exercises
Foot care—dry, frequent cleaning and corn care
Oxygenation to toes/foot, proper shoes (MCR)
O Do not consult **O**thers
Trimming of nail should be done carefully

Remember as **BAREFOOT**

Causes of death in diabetic ulcer of the foot

- Ketoacidosis with septicaemia
- Severe electrolyte abnormalities
- Other causes such as silent myocardial infarction.

PRESSURE SORE (PREVENTION AND TREATMENT)

Pressure ulcers or bedsores are a serious and **frustrating complication** for the paralysed, debilitated or comatose patient confined to a bed or wheel chair. These ulcers form when soft **tissue is compressed** between a bony prominence such as the ischium, sacrum or trochanter and a supporting structure such as the bed or wheel chair (Figs 6.32 and 6.33).

Fig. 6.32: Pressure sore

Fig. 6.33: Pressure gangrene—sore over heel

The growing incidence of spinal cord injuries due to automobile accidents and increased numbers of debilitated geriatric patients admitted to hospitals have drawn more attention to the problem of pressure ulcer prevention and treatment. Pressure ulcer is usually the most important factor that delays rehabilitation of the paraplegic or quadriplegic patient.

Factors predisposing to formation of pressure sore

Most important factor is **pressure**, other factors being **paralysis, paresis, shearing forces, malnutrition, anaemia, advanced age and infection**. Lack of protective sensation in comatose, debilitated patients, prevents them from changing posture. The localised pressure continues and skin ulcer develops. Initially there is tissue anoxia and cell death. Later active inflammation and vasodilatation occurs resulting in reactive hyperaemia. If pressure is removed allowing tissue

[1]Remember it is the patient's leg that may have to be amputated and not the neighbour's!

perfusion and thus wash out of toxic byproducts, initial damage may be reversible. If not, permanent damage will occur. This can happen in one to six hours.

Clinical features

1. Early superficial ulceration

- Erythema, oedema and punctate haemorrhage. Moist irregular ulceration with surrounding erythematous halo.

2. Late superficial ulceration

- Full thickness skin ulceration
- Spreading necrosis of subcutaneous tissue
- Deep inflammatory response spreads in cone-shaped fashion to deeper tissues.

3. Early deep ulceration

- Cicatrization of rolled ulcer edges
- **Eschar** at base of ulcer
- Spread of inflammation and bacterial invasion

4. Late deep ulceration

- Breakdown of fascial plane
- Chronic inflammation and fibrosis of deep tissue (bursa formation)

There is no such thing as a small pressure ulcer. The visible skin wound is merely the "**tip of the iceberg**". 70% of the ulcer is below the skin. Pressure is transmitted in a cone-shaped or pyramidal manner from the skin through each layer of tissue to the bony prominence, so that a cone of tissue destruction is created. The point of the cone is at the skin surface, and base is formed by the larger undermined defect overlying the bone.

Preventive measures

Pressure ulcers can be avoided by meticulous skin care and relief of pressure over bony prominence (Key Box 6.11).

KEY BOX 6.11

SKIN CARE

1. Regular, periodical skin inspection, especially over the bony prominences.
2. Any sign of redness, irritation or abrasion—if noted—***all pressure must be taken off*** the area immediately.
3. Keeping the skin clean and dry: Moist areas lead to maturation. Fine talcum powder may be applied to areas where moisture tends to develop. It must be dusted every day after drying the skin.
4. Gentle massage of vulnerable skin with lanolin lotion.
5. ***Care of perineum and genitalia,*** especially in patients with incontinence.
6. Clothing and bedding must be wrinkle-free, made of porous absorbent material to allow air circulation and avoid accumulation of perspiration.
7. ***Pressure relief***—in bedridden patients.
   a. Frequent ***change of posture*** round the clock every 2 hours.
   b. ***Avoid localised pressure*** by proper body alignment.
   c. ***Use of air or fluid-filled floatation*** mattresses also lessens risk of ulcer formation.
   d. Patient and patient's family education.

Treatment

1. **Superficial ulceration:** Debridement and allowing it to heal by secondary intention. It will take many weeks to heal.
2. **Deep ulceration** or large superficial ulceration
   - Bedside debridement of obviously necrotic material
   - **'Wet to dry' dressing**
   - Use of **desloughing agents**
   - Systemic **antibiotics**
   - **Nutritional** consideration
   - Correction of spasm and contractures, if present.
   - Once it is ready, the defect is closed.

Methods of closure

a. Primary closure—undermine and approximate the cut edges
b. SSG—in selected cases only
c. Skin flaps
   - Transposition flap
   - Rotation flap
   - Advancement flap
d. **Cultured** muscle interposition for severe and ischial pressure sores.

3. **Education of patient** and patient attenders to prevent pressure sores.

MISCELLANEOUS

MAGGOT THERAPY

- A **maggot** is the larva of a fly such as house fly. In practice, any neglected wound on the body specially legs can get contaminated with maggots. Within 1–2 days, there can be acolony of maggots (Fig. 6.34).
- It is a type of biotherapy involving introduction of live, disinfected maggots into nonhealing wounds.
- The ugly look and feeling of a crawling creature on the body drives the patient to reach the hospital in our country. Maggots can be removed by using turpentine.

Mode of action:

- Debride wounds by dissolving necrotic, infected tissue
- Disinfection of the wound by killing bacteria and healing—doubtful
- Maggot therapy may also reduce the need for antibiotics in people with complex, chronic wounds.

Fig. 6.34: Maggots in diabetic patient who had venous ulcer

- Maggot therapy has been shown to accelerate debridement of necrotic wounds.

Indications

- Maggots inhibit and destroy a wide range of pathogenic bacteria including methicillin-resistant *Staphylococcus aureus* (MRSA), group A and B streptococci, and gram-positive aerobic and anaerobic strains. Maggot therapy therefore represents a cost-effective method for managing MRSA infection.
- Maggot therapy may also reduce the need for antibiotics i people with complex, chronic wounds.
- Any chronic ulcers—not responding to conventiona treatment.

DIABETIC ULCER LEG AND RESEARCH

Aldose reductase is the first enzyme in the sorbitol-aldos reductase pathway responsible for the reduction of glucose t sorbitol, as well as the reduction of galactose to galactitol Too much sorbitol trapped in retinal cells, the cells of the lens and the Schwann cells that myelinate peripheral nerves ca damage these cells, leading to retinopathy, cataracts an peripheral neuropathy, respectively. Aldose reductas inhibitors, are currently being investigated as a way to preven or delay these complications.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- More details have been written about pathogenesis and management of diabetic foot. Dozens of varieties of wound dressings are available in the market in case of nonhealing ulcers/wounds, a list of which has been included.
- Remember Eusol and hydrogen peroxide are not favoured nowadays. Hydrogen peroxide is cytotoxic to fibroblasts.
- Povidone iodine is also toxic to fibroblasts. It increases risk of wound infection.
- VAC therapy is added
- Maggot therapy is added

MULTIPLE CHOICE QUESTIONS

1. Which of the following statement is false in tuberculous ulcers?
A. Undermined edges
B. Edge is thin and bluish in colour
C. Apple jelly granulation tissue
D. Wash leather slough

2. Following are true for squamous cell carcinoma *except*:
A. Keratin pearls are diagnostic
B. Induration at the base is significant
C. Ulcers will have everted edges
D. Keratoacanthoma has totally different cytological features than squamous cell carcinoma

3. Following are features of healing ulcer *except*:
A. Serous discharge
B. Sloping edge
C. Slough is absent
D. Signs of inflammation are present

4. Following are true for induration *except*:
A. Maximum induration is seen in malignant melanoma
B. Brawny induration is seen in chronic abscess
C. Carcinoma tongue can present as indurated lesion
D. It is overgrowth of fibrous tissue

5. Gummatous ulcer has following features *except*:
A. Commonly seen in the subcutaneous tissues
B. Floor has wash leather slough
C. Healing results in hypertrophic scar
D. It has punched out edges

6. Alginate wound dressing has following advantages *except*:
A. Nonadherent
B. Absorbs exudate
C. It is safe in renal failure cases
D. With silver, it can act against MRSA also

7. Characteristic feature of venous ulcer in the leg is:
A. Deep painful ulcer
B. Superficial ulcer with pigmentation around
C. Penetrating ulcer with visible bone
D. Ulcers on the dorsum of the foot

8. Following are true for usage of platelet derived growth factors in wound dressings *except*:
A. They are used when vascularity is poor
B. They are used in neuropathic ulcers
C. They act through tyrosine kinase receptor
D. They stimulate angiogenesis

9. Following are true for tropical ulcer *except*:
A. It is caused by Vincent's organism
B. Minimal inflammation
C. It is also an example of phagedenic ulcer
D. Metronidazole is helpful

10. Following are true about sorbitol *except*:
A. Nerve damage in diabetic ulcer foot is due to sorbitol
B. It results from reduction of glucose
C. It occurs when hydroxyl group is changed to aldehyde
D. It can be used as laxative

11. Infrainguinal bypass surgery for diabetic ulcer—following are true *except*:
A. Long saphenous vein is inferior to PTFE graft
B. Preoperative vein marking is helpful
C. 2 years patency is around 70%
D. Amputation rate after surgery is still high

12. Following are true for cilostazol *except*:
A. It reduces the pain of intermittent claudication
B. It is a phosphodiesterase inhibitor
C. Action is similar to pentoxyphylline
D. Dose is 50 mg twice daily

ANSWERS

1 D 2 D 3 D 4 A 5 C 6 C 7 B 8 A 9 B 10 C 11 A 12 C

7

Lower Limb Ischaemia and Popliteal Aneurysm

- Lower limb ischaemia
- Collateral circulation
- Clinical features
- Clinical examination
- Management
- Differential diagnosis
- Acute arterial occlusion
- Critical limb ischaemia
- Popliteal aneurysm
- Ainhum
- Frostbite
- Reperfusion injuries
- Fat embolism, air embolism
- Fontaine classification
- ICU gangrene
- What is new?/Recent advances

Introduction

Walking is a fundamental human requirement. Peripheral arterial disease (PAD) is a main cause of disability. Majority of the individuals are conscious about chest pain caused by myocardial ischaemia and arrive at a hospital early. However, patients present relatively late to the hospital with lower limb ischaemia (Figs 7.1 and 7.2). The disease, even though benign, is not curable totally, thus causing financial, social and psychological burden to the patient and his relatives.

Fig. 7.1: Atherosclerosis with diabetes resulting in wet gangrene — toe has been amputated

Fig. 7.2: Dry gangrene due to atherosclerotic arterial disease—part is dry, mummified and toe has been amputated

CAUSES OF LOWER LIMB ISCHAEMIA (Table 7.1)

Other rare causes of lower limb ischaemia include popliteal entrapment syndrome and cystic medial degeneration. One should not forget that **diabetes mellitus** is also one of the common causes of peripheral vascular disease in elderly patients. While treating a patient with atherosclerotic disease, care of diabetes is equally important.

Risk factors for peripheral arterial disease (PAD)

- **S**moking: Smoking in any form increases the risk by almost tenfold. It is proportional to 'pack-years' smoked.
- **M**ale (gender): Men are affected 10 years earlier than women.
- **O**ther risk factors: This include hypercholesterolemia (> 200 mg/dl), and hypertriglyceridaemia, hypercoagulable state (polycythaemia) and hyperhomocysteinaemia.
- **K**nown diabetes: Specially type 2 diabetes mellitus—increases the risk by two to four times.

Table 7.1 Causes of lower limb ischaemia

Chronic lower limb ischaemia	Acute lower limb ischaemia
• Atherosclerosis	• Acute thrombosis
• Thromboangiitis Obliterans (TAO)	• Acute embolism
• Collagen vascular disorders	• Trauma to the vessels
• Diabetes (diabetic foot—neuro ischaemic foot)	• Aneurysm

- **E**lderly patients between 60–70 years are vulnerable
- **R**aised blood pressure—hypertension

You can *remember as* **SMOKER**

Hyperhomocysteinaemia

- It increases the risk of developing PAD by 7%
- Homocysteine levels more than 15 µmol/l
- Increased levels cause endothelial injury and leads to vascular inflammation
- Defective gene methylene tetrahydrofolate reductase or MTHFR
- Prevention is by eating foods containing B_6, B_9 and B_{12} and with folate, such as potato, green vegetables, fish, etc.
- It is also a strong risk for myocardial infarction in young patients.

Collateral circulation

- Collateral circulation is present in most of the organs. Hence, even if a major vessel is occluded, the organ can still survive provided collaterals are well-developed.
- Chronic ischaemia caused by TAO or atherosclerosis, allows sufficient time for collaterals to develop. Hence, necrosis or gangrene that occurs is minimised. Thus, limbs often survive (Figs 7.3 and 7.4).
- In acute ischaemia caused by **thrombus** or **embolism**, there is no time for collaterals to develop. This results in gangrene of the limb, in untreated cases.

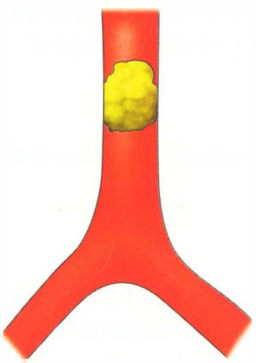


Fig. 7.3: Sudden occlusion. Embolic—no time for collaterals to develop

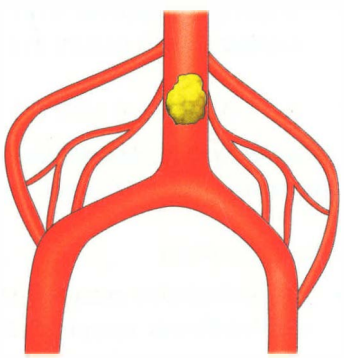


Fig. 7.4: Slow occlusion (atherosclerotic). Collaterals develops

CLINICAL FEATURES—SYMPTOMS

1. **Pain** in the limb is the chief symptom of lower limb ischaemia. It is a severe cramp-like pain, due to ischaemia of the muscles brought on mainly by exertion. It is called **intermittent claudication** (Key Box 7.1).

   Rest pain*[1]*: It is an intractable type of pain usually felt in the foot (instep), toes, etc. It is an indication of severe ischaemia of the foot with impending gangrene. Typically, a patient with rest pain sits on the bed, holds his foot with both hands or may hang the foot out of bed. This gives him some kind of relief. Rest pain is worse at night time[1]. It may lead to suicidal tendency.

   - Claudication distance refers to the distance a patient is able to walk before the onset of pain. A patient with severe claudication may not be able to walk even a few yards.
   - Site of claudication depends upon the level of arterial occlusion (Table 7.2).
   - Also *see* Table 7.3, for other causes of pain in the leg.

2. **Nonhealing ulcer** is the next common presenting symptom. It is usually precipitated by a minor trauma and it occurs in the most distal part of the body such as the tip of toes. Ischaemic ulcers are deep and very painful.

KEY BOX 7.1

GRADES OF INTERMITTENT CLAUDICATION—BOYD'S CLASSIFICATION

Grade I : The patient walks for a distance, gets the pain, continues to walk and the pain disappears. As a result of ischaemia, anaerobic metabolism takes place, which produces ***substance P, lactic acid,*** etc. These produce vasodilatation and the pain disappears.

Grade II: The patient walks for a distance, gets the pain and continues to walk with the pain. He has a limp.

Grade III: The patient walks and gets the pain. He has to take rest. This grade indicates severe muscle ischaemia.

In late stages: Pain at rest is due to ischaemia of nerves* in addition to ischaemia of the muscles.

*Cry of the dying nerves, due to involvement of vasa nervosum

Pain is due to ischaemic neuropathy involving small unmyelinated A delta and C sensory fibres.

Table 7.2

Level of occlusion	Claudication site
Aortoiliac obstruction	Claudication of both gluteal regions, thighs and calves
Iliofemoral obstruction	Claudication of thigh muscles
Femoropopliteal obstruction	Claudication of calf muscles
Popliteal obstruction	Claudication of the foot muscles, **instep claudication**

[1]One of our TAO patients with sleepless nights due to rest pain committed suicide by jumping from the 2nd floor of the hospital.

Table 7.3 Important causes of pain in the leg

Condition	Aetiology	Nature of pain	Location
• Intermittent claudication	Inadequate skeletal muscle perfusion	Burning or cramp like	Calf muscles, thigh or gluteal region. Relieved on rest
• Neurogenic claudication	Lumbosacral nerve root compression	Diffuse radiating deep ache with or without paraesthesia	Pain on first step, decreased on sitting or bending over while walking
• Venous claudication (DVT)	Proximal venous occlusion thrombosis	Bursting	Engorgement during exercise
• Varicosity	Long saphenous varicosity	Diffuse aching/heaviness	Decreases on walking due to calf muscles pump

PEARLS OF WISDOM

Intermittent claudication in a young patient can be due to some rare causes such as
- ***Popliteal artery entrapment due to abnormal origin of gastrocnemius muscle***
- ***Cyst in the media of the popliteal artery***
- ***Hyperhomocysteinaemia.***

3. Some patients present with **gangrenous patches** of skin or subcutaneous tissue. Gangrene affects distal parts such as toes. However, gangrene is minimal because of collaterals (Fig. 7.5).
4. History of **bilateral gluteal claudication** with impotence can occur in a young patient due to a saddle thrombus at the bifurcation of aorta. It is called **Leriche's syndrome**. Impotence is due to failure to achieve an erection due to paralysis of L1 nerve.
   - **Gluteal claudication** is confused for sciatica and many patients are referred to orthopaedic department. Sciatica causes **neurogenic claudication** which is present even **at rest** and is aggravated on movements of the spine. Causes of neurogenic claudication are slipped disc, fracture vertebrae, tuberculosis of spine, etc.
   - Calf muscle claudication (Key Box 7.2)

Fig. 7.5: Thromboangiitis obliterans with dry gangrene

KEY BOX 7.2

SUPERFICIAL FEMORAL ARTERY STENOSIS OR OCCLUSION

- It is the most common cause of intermittent claudication
- Usually calf muscles are affected
- Does not produce life-threatening ischaemia unless profunda femoris is involved.
- Single stenosis less than 3 cm is treated by **P**ercutaneous **T**ransluminal **A**ngiography (**PTA**)

5. Coldness, numbness, paraesthesia and colour changes indicate chronic ischaemia.
6. Majority of patients with peripheral vascular disease are smokers. TAO occurs exclusively in male smokers[1].

PEARLS OF WISDOM

Cauda equina claudication or pseudoclaudication is due to compression of cauda equina.

Classification

- Rutherford also categorised chronic lower limb ischaemia into 6 different categories starting from asymptomatic case to a major tissue loss.
- Claudication has been classified as mild, moderate and severe depending upon treadmill response and ankle pressure (AP)

Mild: Complete treadmill test
AP is > 50 mmHg after exercise

Moderate: Cannot complete treadmill test. AP after exercise is < 50 mmHg

Severe: Resting AP ≤ 40 mm of Hg (cannot do treadmill test)

[1]A 26-year-old female, nonsmoker patient presented to the hospital with ischaemic features of the right upper limb. All causes of upper limb ischaemia were ruled out (Raynaud's, cervical rib, etc.). On careful questioning, she admitted to using SNUFF DIPPING for 10 years (snuff contains nicotine).

SIGNS (CLINICAL EXAMINATION)

Inspection

The ***findings are appreciated*** better if a comparison is made with the opposite limb. Evidence of chronic ischaemia of the leg are:

- ***Attitude of the limb:*** Very often, the patient holds the calf muscles or dorsum of foot.
- Flattening of the terminal pulp spaces of toes
- Fissures, cracks in between the toes
- Ulceration of toes, interdigital ulcers
- Brittle, flat and ridged nails, shiny skin
- Loss of hair and subcutaneous fat
- The limb may appear more dark in dark-skinned patients or markedly pale in fair-skinned patients with vasospastic disease such as TAO (Fig. 7.6).

Fig. 7.6: Attitude of TAO patient—foot is tightly held by hand. Observe gangrene, ulcer, skin changes and ridged nails

- Gangrene is usually dry with a clear line of demarcation. It indicates the junction of dead and living tissue. Since the blood supply to the muscle is better, usually the **line of demarcation involves skin and subcutaneous tissue**. Line of demarcation is very well appreciated in senile gangrene **where it can be skin, muscle or bone-deep**.
- The limb may show atrophy of muscles.
- Multiple toes and finger involvement suggest vasculitis (Fig. 7.7).

PEARLS OF WISDOM

Why is ischaemic pain more in the night?
Loss of gravity assistance to arterial supply, reduction in cardiac output at rest, reactive dilatation of skin vessels to warmth and increased attention of the patient to the leg.

Fig. 7.7: Vasculitis with collagen vascular disorder. It had affected all the ten toes

Palpation

1. **Ulcer:** Examination should be done as described in Chapter 6. Ischaemic ulcers are very tender.
2. **Gangrene:** It is described according to its size, shape and extent. In dry gangrene, the part is dry and mummified or shrunken. Features of dry gangrene are summarised in Key Box 7.3.

KEY BOX 7.3

GANGRENE

- Loss of temperature
- Loss of sensation
- Loss of function
- Loss of pulsation
- Loss of colour

3. **Limb** above[1]
   - **Ischaemic limb is cold:** Careful palpation from above downwards will reveal the change in temperature from warm to the cold area. Temperature changes are appreciated better with the dorsum of the hand which is more sensitive as it has a lot of cutaneous nerve endings.
   - **Tenderness:** It is tender due to the presence of inflammation.
   - **Sensation:** Ischaemic limb is hypersensitive[2], due to the irritation of nerve endings.
   - **Pitting oedema** can be due to thrombophlebitis or due to nonfunctioning of limb.
   - Feel pulses in all four limbs and head and neck.

[1]It is better to palpate the entire limb from the thigh downwards.
[2]Ischaemic limb is like irritable personalities.

4. Palpation of pulses[1] (Table 7.4).

- After examining the pulses, results are interpreted in **a pulse chart** as shown in Key Box 7.4.
- In a similar manner, upper limb pulses, head and neck pulses are also recorded in the pulse chart.

KEY BOX 7.4

PULSE CHART

(For example, classical case of TAO left lower limb)

LOWER LIMB PULSES	Right	Left
Dorsalis pedis	++	-
Posterior tibial	++	-
Popliteal	++	+
Femoral	++	++

++ : Normal; + : Weak; - : Absent.

+++ : Indicates prominent pulse such as water hammer pulse as in aortic insufficiency

Disappearing pulse: When collateral circulation is very good peripheral pulses may be normal. However, when the patien is asked to exercise, the pulse may disappear. Exercise produces vasodilatation below the obstruction and arteria inflow cannot keep pace with increasing vascular space pressure falls and the pulse disappears.

Other tests of minor importance

1. **Buerger's postural test** is relevant in fair-skinned patients. The patient (supine) is asked to raise his legs vertically upwards keeping the knees straight. In cases of chronic ischaemia, marked pallor develops within 2–3 minutes. The angle at which pallor develops is Buerger's angle of circulatory insufficiency. In ischaemic limb, pallor develops even on elevation of leg up to 15–30 degrees.
2. **Capillary refill test:** Apply pressure over the tip of the terminal pulp space for a few seconds and release the pressure. Rapid return of circulation is observed in normal persons (< 2 seconds).

Table 7.4 Examination of peripheral vessels

Artery	Site where it is felt	Remarks
	Examination of lower limb pulses	
1. **Dorsalis pedis** is the continuation of anterior tibial artery	At the level of ankle joint lateral to extensor hallucis longus. It should not be felt distally where it dips into the plantar space	In 10% of cases, it can be absent
2. **Posterior tibial artery** is a branch of popliteal artery	In between the medial malleolus and medial border of the tendoachilles	For circulation of the foot, any one of these arteries is sufficient
3. **Popliteal artery,** a continuation of femoral artery, extends from the hiatus in adductor magnus to the fibrous arch in soleus. It is about 20 cm long	It is felt in the prone or supine position with knee flexed. It is felt against lower end of femur or against tibial condyles	**The knee is flexed to relax popliteal fascia.** Dorsalis pedis, posterior tibial and popliteal artery are usually not palpable in TAO patients
4. **Femoral artery** is the continuation of external iliac artery	It is felt midway between anterior superior iliac spine and pubic tubercle, just below the inguinal ligament in the upper thigh	Abduction and external rotation of the hip joint may facilitate the palpation in obese patients
	Examination of head and neck vessels	
1. **Subclavian artery** arises from the arch of aorta on the left side and brachiocephalic on the right side	It is felt in the supraclavicular region in the posterior triangle against the first rib	Difficult to feel in obese patients
2. **Common carotid artery** arises from arch of aorta on the left side and from brachiocephalic artery on the right side	It is felt against the carotid tubercle of sixth cervical vertebra (C6) in the carotid triangle (at the upper border of the thyroid cartilage)	Carotid artery bifurcates at the upper border of the lamina of thyroid cartilage (C3 vertebra)
3. **Superficial temporal artery** is the terminal branch of the external carotid artery	It is felt in front of tragus of the ear against the zygoma	This is involved in temporal arteritis, a type of giant cell arteritis

[1]A thorough clinical examination of pulses includes not only lower limb vessels but also head, neck and upper limb vessels.

- The test can also be done in the ischaemic foot by asking the patient to sit up and hang his legs down and observe for colour changes. The time taken for the ischaemic foot to become pink is described as capillary filling time. This is prolonged in an ischaemic foot.

Auscultation (Figs 7.8 and 7.9)

1. Systolic bruit over the femoral artery can be heard in atherosclerotic occlusion of iliofemoral segment, due to turbulence created by the blood flow.
2. Auscultation of the heart to rule out mitral stenosis (mid-diastolic murmur, loud 1st heart sound).

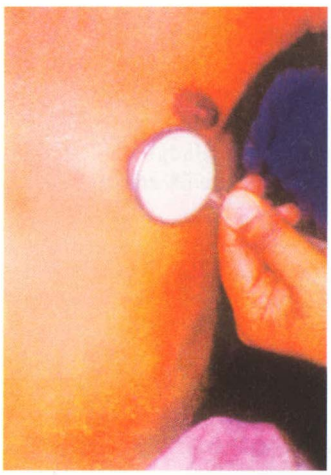


Fig. 7.8: Auscultation of the heart to rule out valvular heart diseases

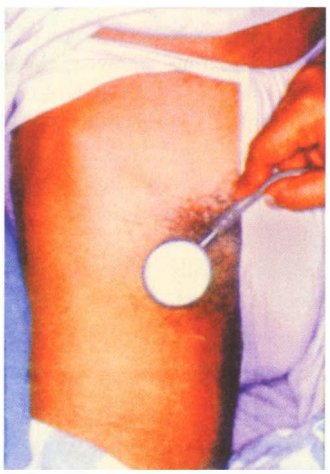


Fig. 7.9: Auscultation over the femoral artery to look for bruit

CLINICAL NOTES

An eighteen-year-old girl was kept in the MBBS examination with ischaemia of lower limb of 6 days duration. The candidate offered collagen vascular disorder as the first diagnosis and failed.

He had missed the cardiac history and findings totally by not auscultating the heart. It was a case of mitral stenosis with acute embolic gangrene of the lower limb.

Differential diagnosis

- Even though there are many causes of lower limb ischaemia, thromboangitis obliterans (TAO) and atherosclerotic vascular disease are the commonest causes. Hence, they have to be considered first before giving other diagnosis.
- TAO is also called Buerger's disease. The details are given in Table 7.5 and Key Boxes 7.5 to 7.7).
- Atherosclerotic vascular disease is the commonest cause of lower limb ischaemia. It can manifest as simple ulcer to massive gangrene.

KEY BOX 7.5

CHECKLIST OF FEATURES OF CHRONIC LOWER LIMB ARTERIAL OCCLUSION

- **C**old and numbness
- **L**imb elevation: Slow venous refilling. Line of demarcation present
- **A**ltered/diminished sensation
- **U**lcerations
- **D**ead toe—gangrene—**D**ried and mummified
- **I**ntractable pain—rest pain
- **C**racks, fissure—interdigital
- **A**rterial pulsations decreased or absent
- **T**hrill/bruit may be present
- **I**ntermittent claudication
- **O**edema—If thrombophlebitis or cellulitis occurs
- **N**arrow calf muscle girth—muscle atrophy

Remember as **CLAUDICATION**

KEY BOX 7.6

CLASSIFICATION OF BUERGER'S DISEASE

Type I :	Upper extremity
Type II :	Crural (leg and foot)
Type III :	Femoral type—femoropopliteal
Type IV :	Aortoiliac
Type V :	Generalised type

KEY BOX 7.7

BUERGER'S DISEASE—SUMMARY

- Male smoker
- Progressive, nonatherosclerotic, segmental, occlusive, inflammatory condition
- ***Occlusion of small- and medium-sized vessels, superficial thrombophlebitis and Raynaud's phenomenon constitute the 'triad' of TAO.***
- Microabscesses, polymorphs, giant cells (pathology) are found.
- Distal, infrapopliteal, segmental occlusion with skip lesions and corkscrew collaterals in angiogram.
- Stop smoking
- Start analgesics
- Lumbar sympathectomy is of some value

Key words can be *remembered as* **PRISON**

Progressive, **I**nflammatory, **S**egmental, **O**cclusive, **N**onathero-sclerotic

Investigation

1. **Complete blood picture:** Anaemia definitely delays wound healing and it also decreases tissue perfusion. High total count indicates secondary infection.
   - Elevated **platelet count** suggests risk of thrombosis.

Table 7.5 Differential diagnosis[1]

	TAO (Buerger's disease)	Atherosclerosis
1. Age 2. Sex 3. Aetiology	20–40 years Exclusively males 1. It is a smoker's disease. Excessive tobacco (nicotine) produces severe vasospasm of the vessels. 2. Excessive smoking produces increased levels of carboxyhaemoglobin which damages these vessels. 3. Low socioeconomic group, recurrent trauma to the foot, poor hygiene are additional factors. 4. Hypercoagulable state 5. Autonomic hyperactivity 6. Autoimmune factors	Around 50 years and above Females are also affected 1. Atherosclerosis is a rich man's disease, who is usually a smoker, diabetic and hypertensive. 2. Strong family history is also present in a few cases. 3. Consumption of high fat diet leading to obesity, lack of regular exercises and hypercholesterolaemia are other factors.
4. **Pathology**	Diffuse inflammatory reaction involving all three coats of vessel **(panarteritis)** causing a thrombus, resulting in occlusion of lumen **(obliterans)**. Polymorphs, giant cells and **micro-abscesses** are found within the thrombus. In severe cases, vein and nerve are bound by fibrous tissue.	Deposition of lipid-rich atheromatous plaque in the intima is the hallmark of atherosclerosis. Plaques tend to be more in **lower abdominal aorta**, coronary arteries, etc. Plaques may undergo **calcification**, ulceration and thrombosis, **dislodge** cholesterol emboli or may weaken the media and produce **aneurysm**.
5. **Vessels involved**	**Small- and medium-sized vessels** such as dorsalis pedis, posterior tibial, popliteal are commonly involved.	Medium-sized and large vessels such as aorta, common iliac, femoral, common carotid, arteries are involved.
6. Upper limb involvement	Not uncommon	Rare
7. Nature of vessel wall	Not thickened	Thickened
8. Blood pressure	Normal in the normal limb and low in diseased limb.	Hypertension is commonly present.
9. **Superficial[2] migrating thrombophlebitis**	Seen in about 30% of cases of TAO. Veins of lower limb are involved and are tender and thickened.	Not seen
10. **Raynaud's phenomenon**	Can be present	Not seen
11. Auscultation—femoral artery	Bruit is not heard.	Bruit can be present as in aortoiliac disease.
12. Angiography	**Cork-screw pattern** of vessels	Shows site of block

- Fasting blood glucose or and **glycosylated haemoglobin** HbA_{1C} is important test as it reflects the duration of diabetes.
- Increased creatinine indicate renal disease.

2. Lipids: Fasting total cholesterol, high density lipoprotein, low-density lipoprotein, and triglyceride concentration—hyperlipidaemia should be controlled to prevent progression of peripheral arterial disease and death from coronary artery diseases.

3. Hypercoagulable status:

- Protein C deficiency is identified as a risk factor for arterial thrombosis especially in patients who will be treated with heparin.
- Heparin induced platelet aggregation and heparin induced thrombocytopaenia
- Antiphospholipid antibody (APLA) syndrome (APLS) is also called Hughes syndrome. It is an autoimmune hypercoagulable state resulting in thrombosis of veins (deep vein thrombosis), thrombosis of artery (stroke) and pregnancy related complications. Treated by aspirin and heparin.

4. Homocysteine levels more than 15 μmol/l

5. Doppler ultrasound blood flow detector:

This test is based on Doppler principle. An ultrasound signal is beamed at an artery and the reflected beam is picked up by a receiver. Frequency changes of the beam due to moving

[1]From MBBS examination point of view, TAO and atherosclerotic vascular disease are to be differentiated.
[2]Migrating thrombophlebitis is also seen in pancreatic malignancy where it is called Trousseau's sign.

Fig. 7.10: Doppler probe over the femoral artery

Fig. 7.11: Handheld Doppler checking the dorsalis pedis artery

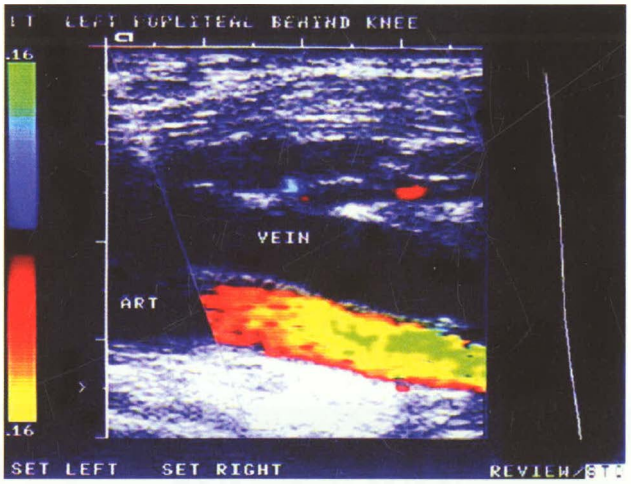


Fig. 7.12: Colour Doppler showing femoral artery and vein

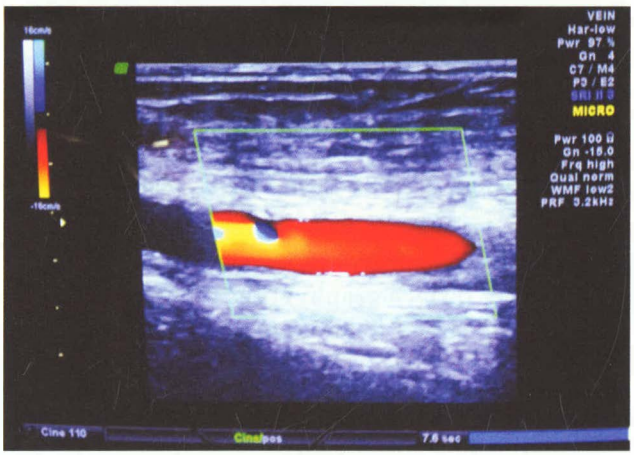


Fig. 7.13: Colour Doppler showing atherosclerotic narrowing

blood are converted into audio signals which can be heard by using a probe. Thus, Doppler probe can be used to detect the pulse even when the pulse is clinically not palpable (Figs 7.10 to 7.13).

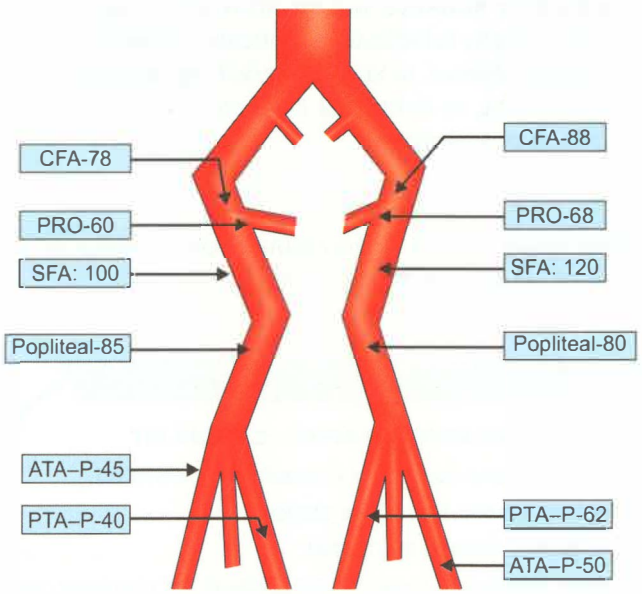
- By using sphygmomanometer, systolic blood pressure (SBP) of the limb can be measured by positioning the cuff at a suitable level and pressure index can be calculated. This is called ankle brachial index—ABI.

$$\textbf{Ankle Brachial Pressure Index} = \frac{\text{Ankle blood pressure}}{\text{Brachial blood pressure}}$$

- Normal values are above 1. However, in patients with peripheral vascular disease of the lower limb, the values are below 1 which indicate vascular obstruction.
- When ankle pressure is less than 30 mmHg, gangrene may be imminent.

Uses of Doppler probe

- To detect normal pulses as in operation theatres.
- To detect clinically nondetectable pulse as in peripheral vascular disease.



CFA—Common femoral artery; PRO—profunda; SFA—Superficial femoral artery; ATA—anterior tibial artery; PTA—posterior tibial artery

Fig. 7.14: Duplex scan report of a patient with the PAD—The number represents the velocity of blood flow in that artery

- To measure BP (blood pressure) in ischaemic limb.
- Remeasuring BP in lower limb after exercise to differentiate ischaemic claudication from neurogenic claudication.

6. Duplex scan:

- This is the investigation of choice today. Duplex scan is a combination of Doppler with B mode ultrasound (Fig. 7.14). With the availability of colour Duplex, the direction of blood flow can be assessed. Red colour means direction of flow towards transducer and blue means away (Key Box 7.8).

KEY BOX 7.8

DUPLEX IMAGING SCAN

- It yields both anatomic and blood flow information
- No nephrotoxic contrast agent is used
- It gives a **triphasic** wave pattern systolic, diastolic and elastic recoil
- Elastic recoil is absent in calcified arteries
- Thus biphasic and monophasic wave patterns indicate 'PAD'
- Overall sensitivity of 92% and specificity of 99% in occlusive cases
- Limiting factors are extensive ulcers, calcification and oedema of leg

7. Angiography (arteriography) (Fig. 7.15)

- Before angiogram, follow certain principles (Key Box 7.9) or precautions.
- It is not usually indicated in TAO patients where direct arterial surgery is not done. However, a few cases of presenile atherosclerosis who are misdiagnosed as TAO will require angiography to locate the site of obstruction which is suitable for an arterial reconstruction. Angiography is indicated in patients with atherosclerotic vascular disease, to know the exact site of block, type of obstruction, to define the collaterals, so as to plan for arterial reconstruction.

PEARLS OF WISDOM

Small vessel bypass is becoming popular today as in selected cases of diabetes and atherosclerosis.

KEY BOX 7.9

PREANGIOGRAPHY—CHECKLIST

- **L**actic acidosis can occur if creatinine is elevated
- **A**nticoagulants: Should be stopped
- **C**reatinine: Should be normal
- **T**ablet Metformin: It can cause acidosis with contrast dye
- **I**ntravenous fluids: To prevent renal failure
- **C**ontrast allergy: Can cause anaphylaxis

Remember as **LACTIC**

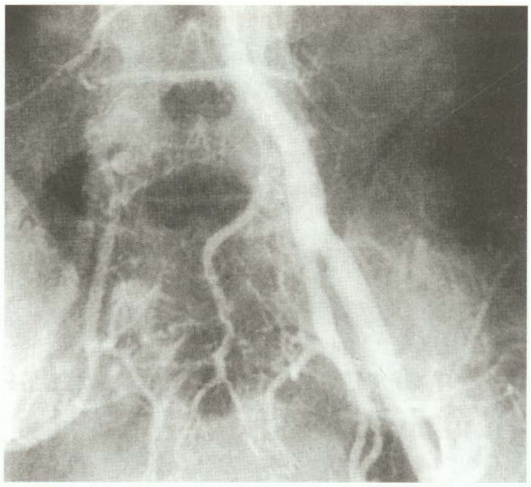


Fig. 7.15: Angiography showing block in the common iliac artery on the right side (conventional)

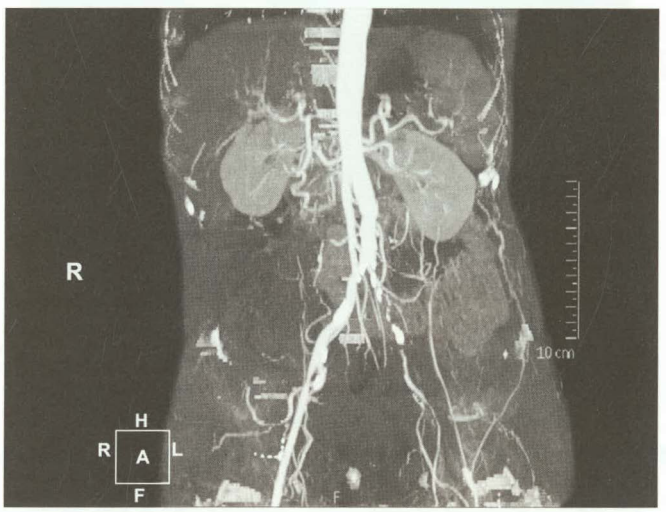


Fig. 7.16: Left common iliac artery occlusion

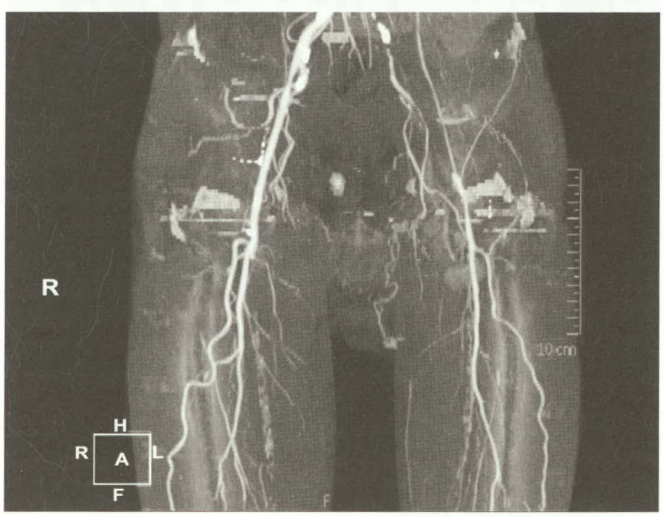


Fig. 7.17: Distal formation at common femoral artery *(Courtesy:* Dr Chandrakanth Shetty, Prof of Radiology and Imaging, KMC, Manipal)

Types of angiography

A. Percutaneous transfemoral retrograde angiography

- This is done in unilateral obstruction. An incision is made in the upper thigh to expose the femoral artery on the normal[1] side. A Seldinger needle and guide wire are used to introduce arterial catheter and radiopaque dye is introduced after placing the catheter into the aorta. It visualises the entire aortoiliac segment and below.

B. Direct translumbar angiography or aortography

- It is indicated when obstruction is bilateral, both femoral pulses are not palpable, clinically manifesting as bilateral lower limb ischaemia. Aorta is directly punctured from behind (translumbar) by using ultrasound image intensifier.

  Results: Arteriography establishes the site of block and nature of collaterals.

Complications of angiography[2] (Key Box 7.10)

- **Thrombosis** at the puncture site resulting in ischaemia.
- **Haemorrhage** from the puncture site which needs to be stopped by pressure packing.
- **Arterial dissection** if catheter is wrongly placed and advanced.
- **Anaphylaxis** can be avoided by a trial injection.
- **Paraplegia** due to spasm of spinal arteries
- **Infection: Digital subtraction angiography (DSA)—** This can be done by arterial or venous route. The arterial route is preferred.

  DSA gives **excellent pictures in carotid and large central vessels**. However, the peripheral vessels may not reveal adequate information. Thus, conventional arteriography still has an important role in atherosclerotic vascular disease.

8. Magnetic Resonance Angiography (MRA) (Figs 7.16 and 7.17)

- It is more popular than arteriography because no arterial puncture and no contrast induced nephropathy.
- Gadolinium enhanced MRA can visualise entire arterial tree pattern including small pedal vessels.
- Patients with newly placed metallic implants are not the candidates.

9. CT Angiography: It is good for above knee vessels compared to below knee (Figs 7.18 and 7.19).

10. Carbon dioxide angiography with gas infusion also has been found useful in all sized vessels and is well tolerated.

KEY BOX 7.10

COMPLICATIONS OF ANGIOGRAPHY

Thrombus
Rarely paraplegia
Arterial dissection
Unexpected infection/sepsis
Massive bleeding
Anaphylaxis
Remember as **TRAUMA**

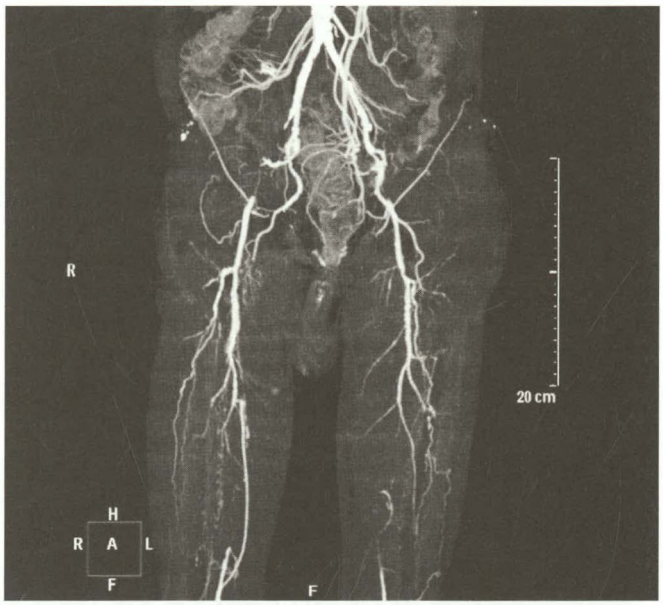


Fig. 7.18: Right common iliac artery occlusion

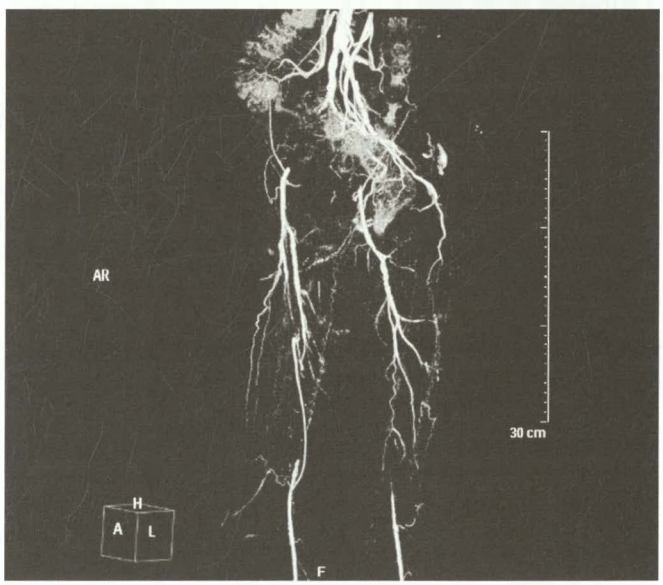


Fig. 7.19: Right common iliac artery occlusion—another view

[1]Normal limb is selected for two reasons:
- Femoral pulse is palpable. Hence, easy to locate the artery and introduce the catheter.
- If a thrombus occurs in the diseased limb due to angiography, it will worsen the ischaemia, may lead to gangrene.

[2]Because of these complications, angiography should be done carefully only when it is indicated.

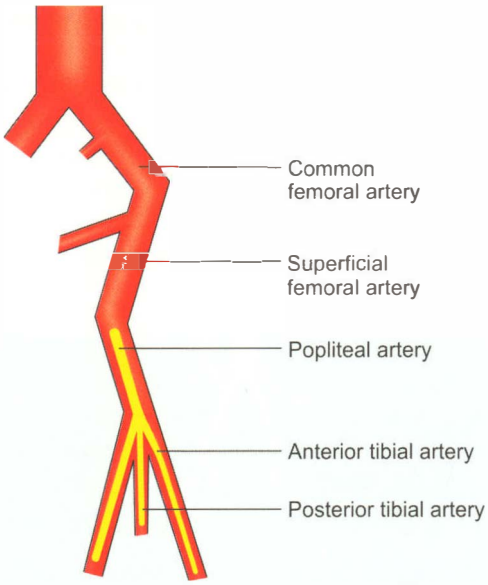


Fig. 7.20: Extensive narrowing of popliteal artery and branches due to atherosclerosis

TREATMENT OF PERIPHERAL VASCULAR DISEASE (TAO AND ATHEROSCLEROSIS)

- In all patients with peripheral vascular disease, following general measures must be taken which will help in better perfusion of the lower limb tissues.
- Anaemia must be treated with haematinics and if necessary, blood transfusion. If ejection fraction is low, drugs are given to improve cardiac output.

Principles (Key Box 7.11)

KEY BOX 7.11

PRINCIPLES OF TREATMENT

- To relieve the pain
- To arrest the progression of the disease
- Role of vasodilators
- Chemical sympathectomy
- Surgical methods

I. To relieve the pain

As already discussed, the pain is very severe and distressing. Some amount of pain relief can be obtained as follows:

1. **Analgesics:** Simple analgesics may not help these patients.
   - **Tramadol** (50 mg) one tablet, 3 times a day can be used.
   - **Ketorolac** 10–20 mg one tablet, 3 times a day can be given in severe cases.
   - Narcotic analgesics can be used judiciously in cases with rest pain.
2. **Buerger's position** by elevating head-end of the bed, causes venous congestion and reflex vasodilatation.
3. **Buerger's exercises** by elevation and dependency of the limb for a few minutes.
4. **Heel raise** by raising the heels of shoes by 1–2 cm, claudication distance can be increased as it decreases work load on the calf muscles.

II. To arrest the progression of the disease

- **Stop smoking:** This is more beneficial in TAO patients than atherosclerotic patients.
- Regular exercise reduces obesity, controls hypertension (Table 7.6).
- **Diet:** Avoid fatty food to reduce serum cholesterol. It is more useful in patients with hyperlipidaemia.
- Avoid injuries.

III. Medical management (Table 7.6)

- Vasodilators have not been found to be useful in atherosclerotic vascular disease.
- Some degree of reduction of pain and healing of cutaneous ulcers has been found in TAO patients.
- The drugs are pentoxifylline and prostacyclin.

Pentoxifylline

- It can be used to treat intermittent claudication
- Pentoxifylline improves blood flow through peripheral blood vessels. It can also be used to increase sperm motility

Table 7.6 Medical management of peripheral vascular disease—drugs, dosage and their role

• Antiplatelet	Aspirin 75 mg with or without clopidogrel 75 mg	Decreases vascular death by 25%
• Antismoking	Stop smoking	Smoking cessation can decrease 10-year mortality rate from 54% down to 18%
• Anticholesterol	Statins—to decrease LDL cholesterol to at least 100 mg/dl	Slow release niacin is emerging as an important therapy in patients with dyslipidaemia
• Antihypertensives	Decrease blood pressure to < 130/85 mmHg	ACE inhibitors, β-blockers
• Antidiabetes	Glycosylated haemoglobin level of < 7%	Oral hypoglycaemic agents or insulin
• Antivasospasm (vasodilator)	Cilostazol—50 mg twice a day (doubtful value)	Inhibits platelet aggregation and is a direct arterial vasodilator

when viable sperms are immotile and are being used in intracytoplasmic sperm injection (ICSI).

- Pentoxifylline improves red blood cell deformability (alters the shape of RBCs) reduces blood viscosity and decreases the potential for platelet aggregation and thrombus formation and reduces inflammation and innate immunity.
- Dose : 400 mg 3 times a day
- It may take 4–8 weeks for the benefit to the patient

Prostacyclins

- **Prostacyclin** (also called **prostaglandin I_2** or **PGI_2**) is a prostaglandin member of the family of lipid molecules known as eicosanoids.
- It inhibits platelet activation and is also an effective vasodilator.
- Prostacyclin (PGI_2) chiefly prevents formation of the platelet plug involved in primary hemostasis (a part of blood clot formation).
- It does this by inhibiting platelet activation.
- It is also an effective vasodilator. Hence used in peripheral arterial diseases.
- Dose: 2 ng/kg/min IV, to start with and increased by 2 ng/kg/min every 15 minutes as tolerated to a maximum of 16 ng/kg/min intravenous.

IV. Chemical sympathectomy

- It acts by producing vasodilatation of the blood vessels of the lower limb.
- In this procedure, 5 ml of phenol in water is injected beside the bodies of 2nd, 3rd and 4th lumbar vertebrae.
- The effect of the drug can be judged immediately by feeling for warmth in the feet.
- It helps in healing of ulcers and may improve rest pain probably by interfering with afferent sensory circuits.

Precautions

1. **Lateral injection** by using a lumbar puncture needle.
2. Injection should be **in front of lumbar fascia** which contains the sympathetic trunk.
3. **Avoid injuries** to aorta and inferior vena cava.
4. Procedure is to be done **under X-ray control** (screening)

Since phenol has replaced lignocaine because of its long lasting effect, it is also called **phenol sympathectomy**.

V. Surgical procedures in TAO

1. **Lumbar sympathectomy** is the indirect surgery done for TAO patients since direct arterial surgery is not possible (Key Box 7.12).
   - **Indications: Cutaneous ulcer and rest pain.**
   - **Structures which can be confused** for lumbar sympathetic trunk are:
     1. Genitofemoral nerve
     2. Tendon strip of psoas muscle
     3. Lymphatic chain and fatty tissue

KEY BOX 7.12

SALIENT FEATURES OF LUMBAR SYMPATHECTOMY

- Transverse loin incision
- Extraperitoneal approach, and it is a preganglionic sympathectomy.
- Lumbar sympathetic trunk is identified in the paravertebral gutter lateral to the psoas muscle as a cord-like structure.
- **2nd lumbar ganglion is large** and has white rami joining it.
- Sympathetic trunk is divided below the first lumbar vertebra and removed up to the 4th lumbar vertebra.
- This is a **preganglionic sympathectomy** because fibres supplying the vessels of the limb have their cell stations in the sacral ganglia which are not disturbed.

   - By depriving the sympathetic nerve supply to lower limb blood vessels, vasomotor tone is reduced so that some amount of vasospasm is reduced. Thus, rest pain improves, minor ulcerations heal due to cutaneous vasodilatation. However, the duration of effect of lumbar sympathectomy is not clear.
   - Both sides can be done in one sitting. However, during bilateral operation, the 1st lumbar ganglion on one side should be spared since removal of both ganglia may cause sterility due to paralysis of ejaculatory mechanism.
   - ***One should be careful not to damage lumbar veins[1] which join inferior vena cava.***
2. **Omentoplasty** has been tried in TAO patients. In most of these patients vasodilator therapy and lumbar sympathectomy has been done with almost no relief of symptoms. In such cases, before a merciful amputation is done, omentoplasty is attempted.
   - By careful division of vascular arcade of omentum, it can be lengthened based on one of the epiploic arteries, brought out of the laparotomy incision, tunnelled in the subcutaneous plane and can be brought up to calf or even to the ankle joint level in some patients.
   - Greater omentum is supposed to produce neovascularisation and thus helps in healing of the cutaneous ulcers. The effect seems to be temporary (not done nowadays).
3. **Conservative amputations** should be done if the toes are gangrenous.
4. **Below knee amputation[2]** is the last resort. It is indicated in severe rest pain cases where all other modalities of treatment have failed. **Risk of amputation after ten years of the disease is around 10%.**

[1]Lumbar veins, if cut accidentally, retract and troublesome bleeding can occur from inferior vena cava. Pressure packing, waiting for 3–5 minutes and then 'see and ligate' should be the policy.

[2]Students are advised not to tell this first as a treatment modality.

KEY BOX 7.13

MAJOR RISK FACTORS FOR ATHEROSCLEROSIS

Lipids: Dyslipidaemia
Inhalation of tobacco: Smoking
Pressure: Hypertension
Insulin deficiency: Diabetes
Disordered metabolism: Hyperhomocystinaemia

Remember as **LIPID**

KEY BOX 7.14

PROGNOSTIC FACTORS FOR LIMB REVASCULARISATION

- Severity of the disease
- Presence of collaterals
- Presence of diabetes
- Chronic smoking
- Site of occlusion
- Age of the patient
- Angina pectoris
- Fitness for anaesthesia

VI. Surgery in atherosclerotic vascular disease

- If possible avoid or treat risk factors (Key Box 7.13).
- The decision to revascularise the limb is taken after an angiography. The success of reconstruction depends upon a number of factors. They are given in Key Box 7.14.
- **Intermittent claudication alone is not an indication for surgery.** Rest pain and pregangrenous changes in the limb are definite indications for reconstruction with accepted mortality and morbidity.
- Surgery can be classified into surgery for aortoiliac disease and surgery for iliofemoral stenotic disease.

Treatment of atherosclerotic disease

I Aortoiliac disease

It is treated by bypass grafts or endarterectomy.

A. Bypass grafts

- Usually, it is bilateral and is treated by using aortobifemoral graft to bypass the stenosis. The graft, made from either teflon or dacron, is used. It is also called Y-graft or trouser graft. It has commonly 16 mm trunk and two 8 mm diameter limbs (Figs 7.21 and 7.22).
- In unilateral cases, unilateral graft is applied.

Figs 7.21 and 7.22: Aortobifemoral graft (*Courtesy:* Dr Ganesh Kamath, Prof. and Head of Cardiothoracic Surgery, KMC, Manipal)

B. Aortoiliac endarterectomy

Indications

- Short segment, large artery such as aorta and single artery.

Types

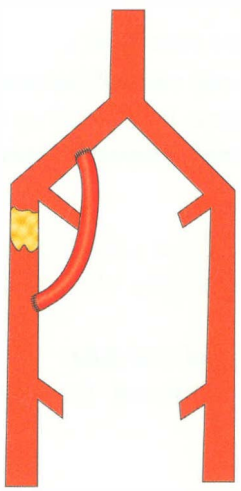
A. Open endarterectomy: An arteriotomy is done first and the diseased intima, atheromatous plaque and the thrombus are removed. An arteriotomy incision can be closed directly or a vein patch graft is used to close the defect, so as to avoid narrowing.

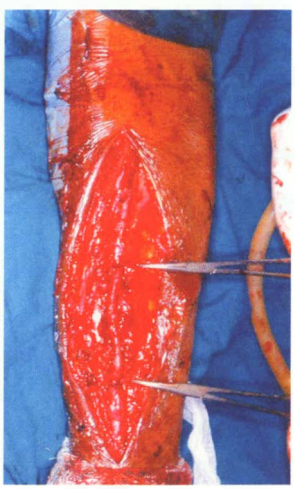
B. Closed endarterectomy is indicated in a longer-diseased segment. In this procedure, after an arteriotomy, a wire loop is used to strip out a core of atheroma by introducing it through the lower arteriotomy and removing the atheromatous plaque from the upper end.

However, results of bypass graft are better than endarterectomy. The modern tendency is to do bypass graft.

II Iliofemoral stenotic disease

- This can be repaired by a **bypass graft** which is sutured to the normal common iliac artery above and to the normal femoral artery below (Figs 7.23 and 7.24).
- If patients are unfit for a major vascular bypass, angioplasty can be done. Ideal for short segment stenosis.
- In this procedure, a **balloon catheter** is inserted into the artery, inflated and its correct position is confirmed by radiopaque markers which are present in the balloon. By inflation and dilatation technique 2–3 times, the stenosed segment can be dilated (Figs 7.25 to 7.31).
- Summary of revascularisation surgery is given in Key Box 7.15).





Figs 7.23 and 7.24: Iliofemoral graft by using reversed saphenous vein

KEY BOX 7.15

SUMMARY OF THE REVASCULARISATION SURGERY (Figs 7.25 to 7.31)

Aortoiliac disease	Aortobifemoral or aortofemoral graft and endarterectomy
Iliofemoral disease	Iliofemoral bypass graft Balloon angioplasty
Femoropopliteal disease	Bypass graft
Profunda artery stenosis	Profundoplasty

- Any arterial stenosis can be dilated in angioplasty.
- The technique can be repeated if stenosis recurs.
- The procedure is done under local anaesthesia and is indicated in poor-risk patients.
- **Ideally suitable for iliofemoral segment**, not suitable for stenosed vessels below knee.
  - ***Internal dissection, distal embolisation, thrombosis and even rupture of vessel can occur.***
  - Recently, angioplasty combined with laser to drill holes through short stenosis have been employed.

III Femoropopliteal occlusion

- This is treated by using a graft extending from the femoral artery above, to the popliteal artery below.
- Reversed long saphenous vein is better than other grafts because it is less thrombogenic.
- Dacron graft, **polytetrafluoroethylene graft (PTFE)**, human umbilical vein graft are the other grafts.

Profunda artery stenosis

- Significant occlusion of profunda is demonstrated by oblique views in an arteriography. If there are no significant vessels available below the stenosis for reconstruction, profundoplasty is considered. It is done by using a patch of Dacron or vein to widen the origin of this vessel after doing endarterectomy.

ACUTE ARTERIAL OCCLUSION

- Sudden occlusion of an artery is commonly due to emboli. The source of emboli is from the heart or from atheroma. Increased incidence of road traffic accidents, fall or war injuries are other causes. Trauma to the artery also produces occlusion.

Embolic occlusion

- This occurs commonly in the peripheral arteries such as the common iliac, femoral and popliteal.
- An embolus is a foreign body to the bloodstream, gets lodged in a vessel and produces obstruction. It manifests clinically as severe ischaemia or gangrene, resulting in **critical limb ischaemia (CLI)**. *See* clinical notes above

CLINICAL NOTES

A 75-year-old lady, known case of polycythaemia vera, presented to the hospital with multiple gangrenous patches of skin in the upper limbs and in the lower limbs. Conservative surgery was attempted by debriding gangrenous portion of the skin. Polycythaemia is also one of the causes of gangrene.

What is critical limb ischaemia (CLI)?

It is defined as persistently recurring ischaemic rest pain requiring regular, adequate analgesia for more than 2 weeks or ulceration or gangrene of foot or toes with an ankle pressure of < 50 mmHg or a toe systolic pressure of < 30 mmHg.

- Atherosclerotic vascular disease, thromboangitis obliterans, acute embolic ischaemia and even diabetes, etc. at some stage can present as CLI.

Pathology (Fig. 7.32)

Causes (*see* Table 7.7)

Clinical features (Key Box 7.16)

- No previous history suggestive of intermittent claudication
- Sudden dramatic symptoms which are described in the form of **5 Ps**
  - **P**ain
  - **P**allor
  - **P**aresis
  - **P**ulselessness
  - **P**araesthesia

1. **Pain** is severe, unbearable, burning or bursting type.
2. Limb is **pale**, cold and superficial veins are collapsed.
3. **Paresis:** Depending on the level of occlusion, the function of the limb is lost. Movement of the toes becomes difficult, followed by total paralysis.
4. **Pulselessness:** Characteristically, peripheral pulses below the level of embolism are not palpable.

KEY BOX 7.16

SIGNS OF ACUTE LOWER LIMB ISCHAEMIA

- **P**eripheries are cold
- **P**allor of the limb
- **P**oor capillary return
- **P**ositive Buerger's test
- **P**rogressive paralysis
- **P**ulses are absent
- **P**ulse at ankle by Doppler—undetectable *Observe* **7 Ps**

BALLOON ANGIOPLASTY IN ATHEROSCLEROTIC ARTERIAL DISEASE

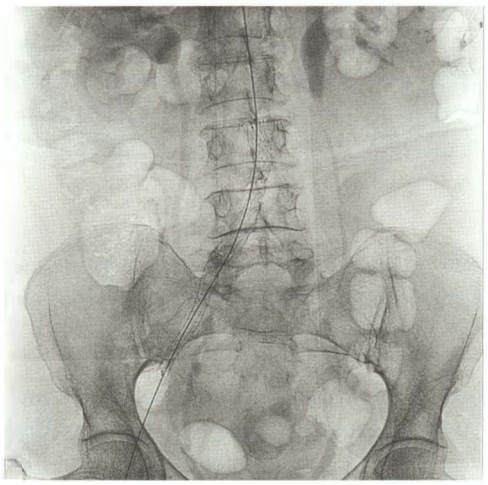


Fig. 7.25: Plain radiograph with guide wire in the aorta

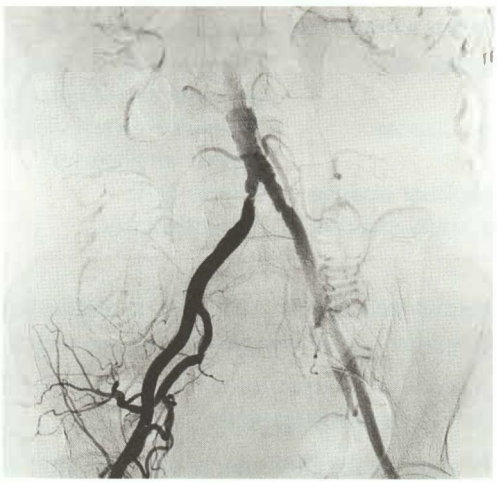


Fig. 7.26: Narrowing of the right common iliac artery

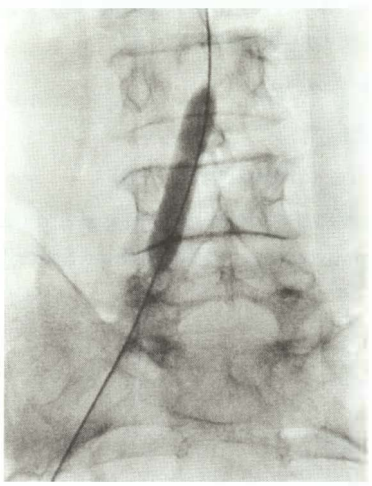


Fig. 7.27: Balloon angioplasty of narrowed right common iliac artery

Fig. 7.28: Post balloon angioplasty

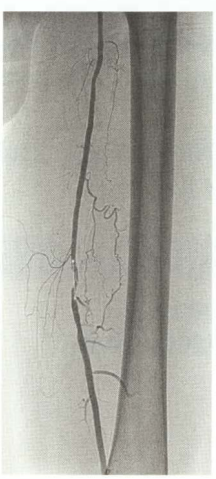


Fig. 7.29: Mid-SFA showing narrowing

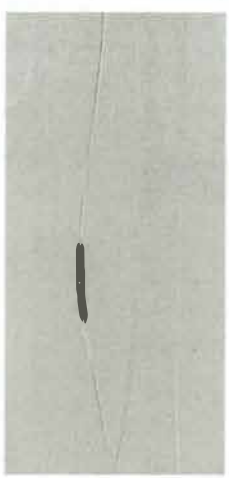


Fig. 7.30: Balloon angioplasty

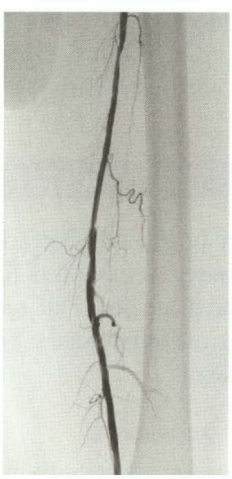


Fig. 7.31: Postangioplasty image showing restoration of lumen of mid-SFA

(*Courtesy:* Prof Chandrakanth Shetty and Dr Praharsha, Resident, Department of Radiodiagnosis and Imaging, KMC, Manipal)

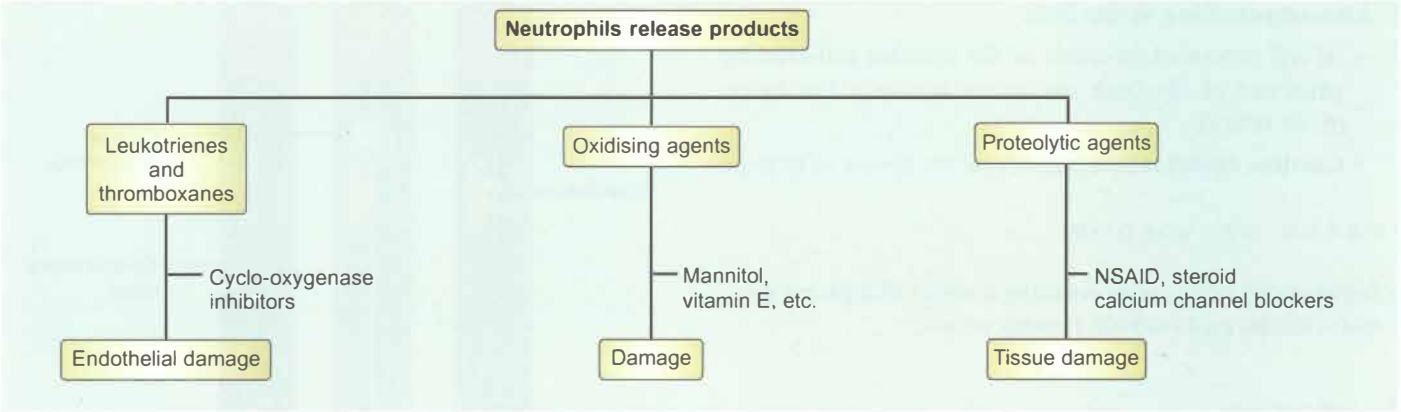


Fig. 7.32: Pathogenesis of tissue damage

Fig. 7.33: Critical limb ischaemia (CLI)

Fig. 7.34: CLI due to diabetes and atherosclerosis

Table 7.7 Causes of acute lower limb ischaemia

Acute lower limb ischaemia

Embolism
- Atrial fibrillation
- Mitral stenosis
- Myocardial infarction
- Aneurysm
- Atheromatous plaques
- Emboli from myxoma of heart

Fig. 7.35

Thrombosis (causes)
- Polycythaemia rubra vera
- Thrombocythaemia
- Leukaemia
- High oestrogen pill
- Atheromatous

Common sites
- Aortic bifurcation (saddle embolus)
- Common femoral bifurcation
- Popliteal trifurcation

Common arteries affected
- External iliac
- Profunda
- Popliteal

- Emboli impact at branching points where arterial lumen narrows abruptly.

5. Altered sensation in the limb.

- If left untreated, necrosis of the muscles followed by gangrene of the limb can occur within a few hours (6–24 hours).
- **Cardiac examination** may reveal the source of emboli.

PEARLS OF WISDOM

Intravenous drug abuse remains a major risk factor for endocarditis and embolic complications.

Investigations

- Peripheral circulation should be assessed by Doppler ultrasound, which is an excellent noninvasive investigation to judge the severity, level, position and length of superficial femoral artery stenosis.
- Digital subtraction angiography in aortoiliac lesions.

Treatment

I. Angioplasty: Percutaneous transluminal angioplasty (PTA) is **indicated in short stenotic lesions in a large vessel**, e.g. iliac and femoropopliteal lesions.

- First, the balloon catheter is introduced percutaneously over a guidewire across the lesion. Under fluoroscopic control, the balloon is dilated until satisfactory widening of the lumen is achieved.
- Relatively safe and simple procedure.
- Immediate intravenous infusion of heparin (10,000 IU) is necessary to reduce the extension of the thrombo-embolism.

II. Emergency embolectomy is done under GA or local anaesthesia either by direct arteriotomy incision and removal of clot or by using a Fogarty balloon catheter to remove an embolus from a vessel remote from arteriotomy (Figs 7.36 and 7.37).

- **Embolectomy:** Under local or general anaesthesia, a transverse incision is given over common femoral artery. A Fogarty catheter is introduced through the incision for about 1–2 cm and the balloon is inflated.

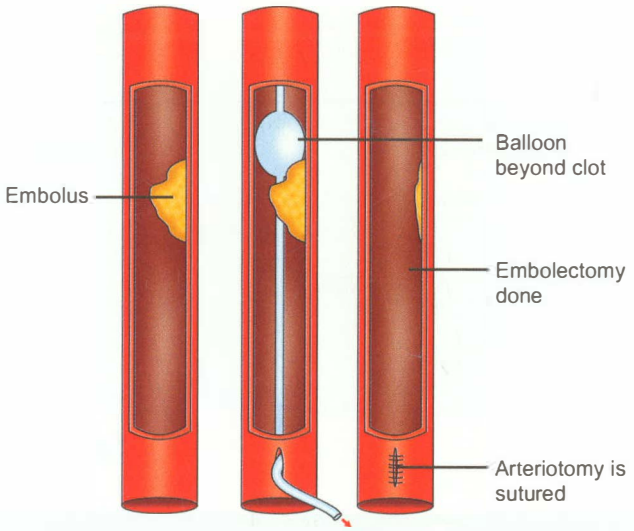


Fig. 7.36: Fogarty embolectomy

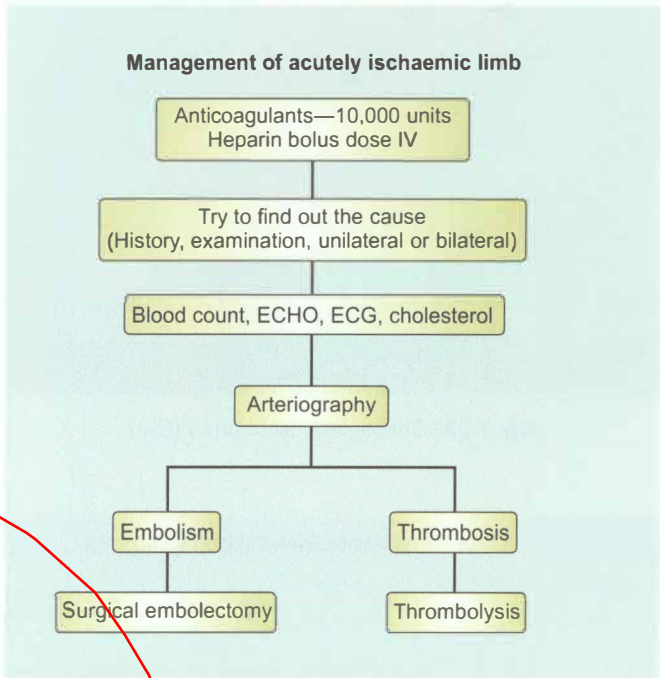


Fig. 7.37: Management of acutely ischaemic limb

Table 7.8 Emboli in various arteries—symptoms, signs and effects

1. Leg	Common femoral, popliteal	Pain/pallor	Gangrene/ischaemia
2. Brain	Middle cerebral artery	Minor or major TIA	Stroke/hemiplegia
3. Retina	Central retinal artery	**Amaurosis fugax**	Fleeting/permanent blindness
4. Intestine	Superior mesenteric artery	Postprandial pain, shock	Intestinal ischaemia/gangrene
5. Kidney	Renal artery	Pain in loin, haematuria	Renal ischaemia
6. Upper limb	Brachial artery	Pain, ulcer upper limb	Gangrene

- The catheter is then withdrawn and emboli removed. The procedure is repeated until bleeding occurs. The catheter can be passed by the ankle and distal emboli also can be removed. Postoperatively, anticoagulants are continued.
- **Intra-arterial thrombolysis:** A catheter is passed into the 'clot' after doing arteriography and ***t-PA*** (***tissue plasminogen activator***) is infused through catheter. Repeat angio check films are taken to see the results such as lysis of clot, revascularisation, etc.

III. Thrombolysis

- It is indicated in acute or acute on chronic ischaemia.
- A fine lysis catheter is passed percutaneously into blocked vessel.
- Streptokinase is infused at a rate of 5000 IU at 10 ml/hr.
- It will reopen the occluded lumen within 24 hours.
- Repeat angiogram is necessary to check for patency.

For contraindications *see* Key Box 7.17.

PERIPHERAL ANEURYSMS

- These can affect the popliteal artery, femoral artery, iliac artery, etc. **70%** of peripheral aneurysms affect popliteal artery, and **two-thirds of them are bilateral**.
- However, students should realise that aneurysms are uncommon (rare) causes of lower limb ischaemia. Early diagnosis and effective treatment are essential to save the limb. **Aortic aneurysms** are discussed in Chapter 37 under abdominal mass.

POPLITEAL ANEURYSMS

They are the **commonest peripheral aneurysms** because of following reasons:

- Turbulence beyond stenosis at the adductor magnus hiatus
- Repeated flexion at the knee.

KEY BOX 7.17

CONTRAINDICATIONS TO THROMBOLYTIC THERAPY

Absolute

- Recent major bleeding
- Recent major surgery
- Recent trauma
- Recent ophthalmologic procedure
- Recent stroke

Relative

- Active peptic ulcer disease
- Pregnancy
- Uncontrolled hypertension
- Coagulation abnormalities

Clinical features

- They affect elderly patients and atherosclerosis is the cause. Age at presentation is 65 years.
- **One-third** of the cases are associated with aortic aneurysm.
- Striking preponderance in **males**. Male:female ratio is 20–30:1.
- Presents as a swelling behind the knee.
- Dull aching pain is common. Severe **bursting pain** indicates **rapid expansion** and impending rupture.
- Pulsatile, tense, cystic, fluctuant swelling behind the knee, in the popliteal fossa and in the line of popliteal artery.
- Its **size diminishes on extending the knee** as the aneurysm is deep to popliteal fascia.
- **Proximal compression test:** On occluding the femoral artery proximally, the swelling may diminish in size.

PEARLS OF WISDOM

In all cases of popliteal aneurysm, please search for femoral and aortic aneurysm.

Investigations

- **Duplex ultrasonography** is the investigation of choice which can measure diameter and determine extent of mural thrombus.
- **Angiography** can demonstrate the extent of involved segment to look for patency and quality of runoff vessels.

Complications

1. **Thrombosis** causes severe acute ischaemia of the lower limb (incidence: 40%).
2. **Embolisation** causes ischaemic ulceration of the lower limb.
3. **Rupture** causes pain and haematoma (rare: 2–5%).
4. **Compression** on the popliteal vein causes pain, tenderness and swelling of the leg.
5. **Compression on the lateral peroneal nerve** causing foot drop, due to paralysis of the peronei and the extensors of the foot.

Treatment

- **Proximal and distal ligation of the artery** followed by reversed saphenous vein bypass graft is the treatment of choice. This method results in total obliteration of the sac with revascularisation of the limb.
- **Excision of the sac is better avoided** because of chances of injury to the popliteal vein and nerves. Lateral popliteal nerve injury causes foot drop and tibial (medial popliteal) nerve injury causes thinning of the calf region, inability to plantar flex the ankle and clawing of the toes due to paralysis of the intrinsic muscles of the foot.

MISCELLANEOUS

AINHUM (Fig. 7.38)

It affects those who do not use footwear or walk barefooted. It starts as a fissure at the level of interphalangeal joint of a toe, usually fifth. Repeated trauma of minor degree may be present. The tissue becomes a fibrous band resulting in tight constriction and necrosis. If it continues, it may culminate in autoamputation.

- The division of band or early Z-plasty may be needed to avoid amputation.
- Thrombolysis with catheter *in situ,* in early cases.

FROSTBITE

- It occurs due to too much of exposure to cold weather.
- High altitudes with excessive cold precipitates vasospasm and damage to the blood vessel wall. It causes sludging of blood and thrombosis.
- Malnutrition, ageing process are the other precipitating factors. Severe burning pain, discolouration of foot, development of blisters suggest gangrene is imminent (Fig. 7.39).

Treatment

- Slow warming of the parts and protection with cotton wool.
- Analgesics and antibiotics.
- Paravertebral injection into the sympathetic chain helps in a few patients.
- Elevation of the foot to reduce oedema.
- Frank cases of frostbite with gangrene require conservative amputation.

REPERFUSION INJURIES OR SYNDROME

- This dangerous event follows revascularisation of limbs, resulting in **acute compartment syndrome** with compartmental pressure exceeding capillary pressure (30 mmHg) (Fig. 7.39).

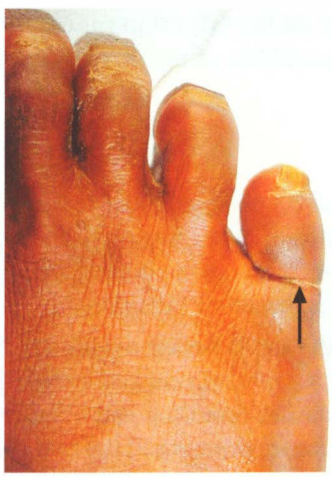


Fig. 7.38: Ainhum

- Most of the injury is believed to be due to O_2 derived free radicals. The most important ones include superoxide radical, hydrogen peroxide and hydroxyl radical.
- These radicals attach unsaturated bonds of fatty acids within the phospholipid membranes resulting in damage.

Management

- Diagnosis is clinical as suggested by severe pain in the limb, oedema of the leg and muscle tenderness.
- Raised intracompartmental pressure measured by transducer cannula will help in the diagnosis.
- Creatinine is elevated suggesting renal failure.
- Creatine kinase will be elevated suggesting rhabdomyolysis.
- Treated by urgent multiple fasciotomy, decompression followed by debridement of dead tissues.

FAT EMBOLISM

Definition

Often a potentially lethal condition which occurs due to blockage of major arteries by aggregation of chylomicrons.

Causes

- Fracture femur (long bones)
- Orthopaedic surgery, multiple fractures
- Liposuction
- Sickle cell disease
- Pancreatitis
- Diabetes mellitus

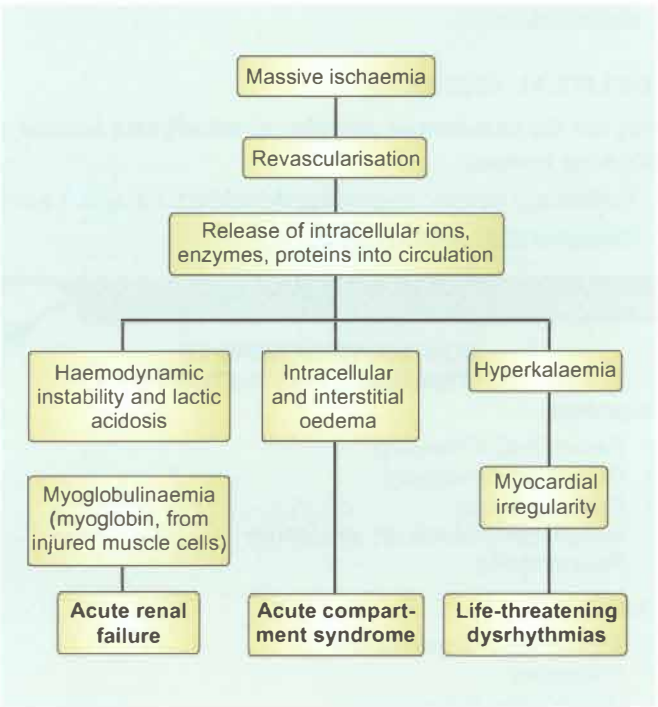


Fig. 7.39: Pathophysiology of reperfusion injuries

PEARLS OF WISDOM

Respiratory failure, confusion, petechial haemorrhages: Triad of fat embolism syndrome. Early diagnosis and prophylactic administration of oxygen to patients at risk is recommended.

Clinical features

- **Pulmonary:** Central cyanosis, tachypnoea, right heart failure, froth in mouth and nostrils. ***Respiratory distress is the most common presenting feature.***
- **CNS:** Drowsiness, disorientation, restlessness, constricted pupils, pyrexia and coma.
- **Retinal artery:** Striate haemorrhages, fluffy exudates on fundoscopic examination, cotton wool spots.
- **Cutaneous:** Petechial rashes in the nondependent parts of the body—chest, axilla and conjunctiva.

Pathogenesis

- 50% of cases occur within 72 hours.
- **Free fatty acids (FFA)** released at the time of trauma or breakdown of fat in the lung directly affect pneumocytes resulting in acute respiratory of distress syndrome **(ARDS)**.
- Other theory is blockage by **aggregation of chylomicrons**.

Investigations

- Arterial blood gases will show hypoxaemia and respiratory alkalosis from hyperventilation.
- Chest X-ray will show bilateral interstitial and alveolar infiltrates in severe cases '**snowstorm**' pattern.

Treatment

- **Close monitoring** with pulse oximetry.
- **Supportive therapy** with oxygen and mechanical ventilation as required.
- **Surgical**—early internal fixation of the fracture.

AIR EMBOLISM

Definition

It is a **potentially fatal condition** that can occur due to blockage of pulmonary artery by **large volume of air** in the venous circulation (Key Box 7.18).

Causes

- **Neck surgery** during which a large vein is inadvertently opened and the patient is in head-elevated position (thyroid surgery) or sitting position (posterior fossa surgery).
- Rapid infusion of intravenous fluids where emptying of the bottle and infusion set may go unnoticed and a fresh bottle is connected and infused without eliminating air from the infusion set.

Effects

Formation of air lock within pulmonary artery and right heart failure.

KEY BOX 7.18

AIR EMBOLISM

- Large veins get opened in the neck
- Open heart surgery or if pulmonary vein is punctured
- Fallopian tube insufflation
- Illegal abortion (through paravertebral veins)
- Paradoxical embolism, reaching coronary artery through patent foramen ovale

Treatment

- Position the patient **head down** (Trendelenburg position) so that air entrainment is stopped.
- Turn the patients on their **left side** so that air will float to ventricular apex reducing its entry into the pulmonary artery.
- Administer **oxygen and resuscitate as necesssary**.

FONTAINE CLASSIFICATION

- Peripheral artery occlusive disease is commonly divided into stages as introduced by Dr René Fontaine in 1954.
- Fontaine classification has defined the severity of chronic ischaemia as:
  Stage 1: Asymptomatic
  Stage 2: Intermittent claudication limiting lifestyle
  Stage 3: Rest pain due to ischaemia
  Stage 4: Ulceration or gangrene due to ischaemia
- Some have also classified **stage 2 into A and B:**
  A – Intermittent claudication > 200 metres
  B – Intermittent claudication for < 200 metres

INTENSIVE CARE UNIT (ICU) GANGRENE

- It has been described previously in literature as a rare clinical entity. It was first described by Jonathan Hutchinson in 1891 as multiple extremity ischemia and was termed 'symmetrical peripheral gangrene'.
- Possible aetiological factors include sepsis, DIC and use of vasopressor agents. The possible responsible drug is noradrenaline (Fig. 7.40).
- Patients in ICU also have multiple arterial punctures for securing intra-arterial line for invasive monitoring in the ICU. The arterial punctures can be a possible aetiological factor.
- Sepsis in the presence of sluggish blood flow has been described to be responsible for development of symmetrical peripheral gangrene (Fig. 7.41).
- Vasospastic conditions, small vessel obstruction and very low cardiac output states (perfusion pressure falling to 35–60 mmHg) may contribute.

Please note: Aortic aneurysm is discussed in Chapter 44 under cardiothoracic surgery.

- Medical conditions such as diabetes mellitus, malignancy, protein C or S or antithrombin III deficiency are also contributing factors.
- The usual manifestation is pallor or cyanosis, coldness and pain in the extremity. They become erythematous and there is dusky discolouration of skin, bullae or blisters followed by gangrene.
- Pulses may be intact in the early stages and large vessels are often spared. Low flow states result in occlusion of the microcirculation of the affected parts.
- The first line of management of this gangrene is immediate discontinuation of vasopressors as soon as discolouration is seen.

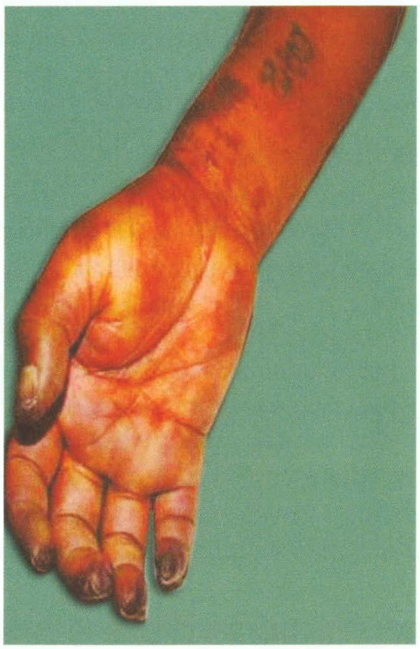


Fig. 7.40: ICU gangrene

Fig. 7.41: Bilateral symmetrical digital gangrene due to atherosclerosis, diabetes, sepsis and hypotension

- Aggressive treatment of sepsis with intravenous antibiotics and anticoagulation for DIC are the suggested measures.
- Local debridement and secondary skin grafting have been unsuccessful.
- Amputation should be considered only after a clear line of demarcation develops. The condition has a high mortality rate of 40%.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- A few clinical photographs and Key Boxes have been added. More details about medical management of arterial disease is added.
- Transluminal angioplasty has been discussed in more detail.
- Fat embolism/air embolism, Fontaine classification are added.
- New photographs, Key Boxes are added.
- ICU gangrene is added.

MULTIPLE CHOICE QUESTIONS

1. Which of the following cannot occur in thrombo-angiitis obliterans?
A. Dorsalis pedis and posterior tibial are often affected
B. Radial artery can also be affected
C. Superficial femoral is also affected
D. Bruit present over femoral artery

2. Following are true for atherosclerotic arterial disease *except:*
A. Large sized arteries are affected
B. Upper limb is often affected
C. It can be associated with aortic aneurysm
D. A bruit can be present over the artery

3. Following are features of thromboangiitis obliterans *except:*
A. Raynaud's phenomenon
B. Migrating thrombophlebitis
C. Segmental panarteritis
D. Polymorphs and giant cells are absent in histopathological examination

4. Following are uses of Doppler probe *except:*
A. Used to feel the nonpalpable pulse
B. To look for pressure index
C. To measure blood pressure
D. To detect triphasic pattern

5. Lumbar sympathectomy has following advantages *except:*
A. Rest pain improves to a small extent
B. Ulcerations heal
C. Claudication and claudication distance improves
D. Nutritive value of the blood flow improves

6. Fogarty catheter is used in:
A. Chronic occlusion of artery
B. Acute embolic occlusion of an artery
C. Vasospastic disease
D. Femoral vein thrombosis

7. Characteristic feature of critical limb ischaemia has one of these:
A. Intermittent claudication
B. Ankle pressure is less than 70 mm of Hg
C. Toe systolic pressure is less than 30 mmHg
D. Absent pulses

8. Triad of fat embolism includes:
A. Respiratory failure, confusion, petechial haemorrhages
B. Respiratory failure, confusion, large purpuric spots
C. Respiratory failure, alertness, petechial haemorrhages
D. Cardiac failure, confusion, petechial haemorrhages

9. Following are true for popliteal aneurysm *except:*
A. It is caused by atherosclerosis
B. It is the most common type of peripheral aneurysm
C. It is always unilateral
D. Foot drop can occur

10. Following are true about lumbar sympathectomy *except:*
A. It is a postganglionic sympathectomy
B. Sympathetic trunk is divided below the first sympathetic ganglion
C. It is a preganglionic sympathectomy
D. Usually done by extraperitoneal approach

11. Infra inguinal bypass surgery for diabetic ulcer—following are true *except:*
A. Long saphenous vein is inferior to PTFE graft
B. Preoperative vein marking is helpful
C. 2 years patency is around 70%
D. Amputation rate after bypass surgery is still high

12. Following are major risk factors for atherosclerosis *except:*
A. Dyslipidaemia
B. Tobacco smoking
C. Hyperhomocystinaemia
D. Alcohol intake

13. Following are true for profundoplasty *except:*
A. It is done by using patch of dacron
B. It is done by using vein graft
C. Done after endarterectomy
D. Done in TAO patients

14. About tissue plasminogen activator, following are true *except:*
A. First purified from melanoma cells
B. It is an endogenous enzyme like urokinase
C. It is sometimes elevated in carcinoma stomach
D. Streptokinase also has plasminogen activating action

ANSWERS

1 D	2 B	3 D	4 D	5 C	6 B	7 C	8 A	9 C	10 A
11 A	12 D	13 D	14 C						

8

Upper Limb Ischaemia and Gangrene

- Raynaud's disease
- Thoracic outlet syndrome
- Axillary vein thrombosis
- Vasculitis syndrome
- Gangrene, cancrum oris
- Acrocyanosis
- Drug abuse and gangrene
- Iatrogenic drug-induced gangrene
- Subclavian steal syndrome
- What is new?/Recent advances

Introduction

Upper limb ischaemia (ULI) is a well recognised clinical entity as lower limb ischaemia, though it is less common. There are certain specific conditions which are responsible for ULI such as cervical rib, Raynaud's disease, etc. It is also important to note that reconstructive surgery is rarely done in the upper limb when compared to lower limb. However, students should be able to identify the ischaemic features in the upper limb early and refer the case to a suitable specialist, so that it can be treated promptly and adequately, thereby avoiding a tragedy such as loss of the limb.

Causes of upper limb ischaemia

1. Raynaud's disease and Raynaud's syndrome
2. Embolic causes
3. Thoracic outlet syndrome (Fig. 8.1)
4. Trauma
5. Buerger's disease
6. Axillary vein thrombosis
7. **Vasculitis syndromes**
   - Takayasu's arteritis, giant cell arteritis, polyarteritis nodosa
   - Systemic sclerosis—scleroderma—**CREST syndrome**

RAYNAUD'S DISEASE (PRIMARY RAYNAUD'S PHENOMENON)

- It occurs in young women, commonly.
- Upper limb is more involved than lower limb

Fig. 8.1: Gangrene of the tip of fingers due to cervical rib

- Commonly seen in western countries in white-skinned people. Cold climate is possibly a precipitating factor. It was first described by Raynaud as **bilateral episodic digital ischaemia of the upper limb** on exposure to cold and emotions. **It is also referred to as primary Raynaud's phenomenon.**
- Raynaud's phenomenon is the blanket term used to describe cold-related digital vasospasm (*see* pathophysiology). Raynaud's phenomenon is subdivided into Raynaud's syndrome where there is an associated disorder and primary Raynaud's disease where there is none.
- **CREST syndrome: C**alcinosis circumscripta, **R**aynaud's phenomenon, (O) **E**sophageal defects, **S**clerodactyly, **T**elangiectasia.

KEY BOX 8.1

SECONDARY RAYNAUD'S PHENOMENON CAUSES

- Atherosclerosis
- Systemic lupus
- Scleroderma
- Cervical rib
- Carpal tunnel syndrome
- Vibrating tools—vibration white finger

- Causes of **Secondary Raynaud's phenomenon** are given in Key Box 8.1.
  Currently, both primary and secondary varieties have been grouped together.

Pathophysiology

On exposure to cold[1], some kind of discomfort and colour changes are observed. This is due to abnormal sensitivity of the arterioles to cold. Three stages have been described.

1. **Stage of syncope:** Arterioles undergo constriction as an abnormal response to cold. As a result of this, the part becomes blanched and severe pallor develops.
2. **Stage of asphyxia:** After a brief period of vasoconstriction, capillaries dilate, filling with deoxygenated blood resulting in bluish discolouration of the part (cyanosis).
3. **Stage of recovery or stage of rubor:** As the attack passes off, relaxation of the arterioles occurs, circulation improves and redness occurs. Because of dilatation of capillaries, red engorgement of the part occurs, which causes tingling, burning or bursting pain in the fingers.

Clinical features

- Affects young women.
- Typically causes bilateral episodic digital ischaemia on exposure to cold.
- **Thumb is usually spared.**
- **Peripheral pulses are normal.**
- Pallor, cyanosis and rubor are the colour changes occurring during the attack along with pain.
- In a few patients, because of **recurrent attacks**, gangrenous patches occur on the tip of the fingers (superficial necrosis).

Differential diagnosis

- Cervical rib
- Vasculitis syndromes
- TAO affecting the upper limb usually affects male smokers. Peripheral pulses are feeble or weak.

[1]Cold refers to temperature—cold climate (winter), cold environment (refrigerator), or cold substance like cold water or ice. Erythromelalgia is a condition wherein heat provokes an attack of burning pain of hands and feet.

Treatment

I. Conservative line of treatment

- Reassurance
- Avoid unnecessary exposure to cold
- Avoid smoking
- Calcium antagonists such as nifedipine 10–20 mg, two times a day may be beneficial.
- If these measures fail, surgery is undertaken.

II. Cervical sympathectomy (Key Boxes 8.2 and 8.3)

- In this operation, sympathetic trunk from the lower half of the stellate ganglion to just below the 3rd thoracic ganglion is removed.
- **Upper ½ of the stellate ganglion is preserved** to avoid Horner's syndrome.
- All rami communicantes associated with the 2nd and 3rd ganglia are removed.
- **Nerve of Kuntz,** a grey ramus which springs from the 2nd thoracic ganglion to the 1st thoracic nerve is also divided.
- Commonly done through a supraclavicular route and axillary route. Thoracoscopic sympathectomy is becoming popular.
- Sympathectomy **raises the threshold** at which spasm occurs (but the effect seems to be temporary). However, the severity of the disease is reduced.

KEY BOX 8.2

INDICATIONS FOR CERVICAL SYMPATHECTOMY

- Raynaud's disease
- TAO
- Hyperhidrosis
- Cervical rib
- Causalgia

KEY BOX 8.3

COMPLICATIONS OF CERVICAL SYMPATHECTOMY

- Perforation of pleura causing pneumothorax
- Lymph fistula due to injury to thoracic duct
- Horner's syndrome
- Injury to the accessory nerve
- Haemorrhage

Other vasospastic disorders

These are rare (Key Box 8.4)

KEY BOX 8.4

OTHER VASOSPASTIC DISORDERS

Acrocyanosis
- Women with cyanosis of hands and feet
- Cutaneous vasoconstriction is the cause

Livedo reticularis
- Spasm of arterioles and dilatation of venules
- Worsening by cold
- May be associated with systemic lupus erythematosus (SLE)

Erythromelalgia
- Burning sensation of hands and feet due to heat

THORACIC OUTLET SYNDROME

SURGICAL ANATOMY OF THORACIC OUTLET

- Thoracic outlet is a tight space with bony structures all around (Key Box 8.5) such as manubrium sternum in the front, spine posteriorly and first rib laterally.
- At the root of the neck, brachial plexus and subclavian artery pass through scalene triangle into the axilla.
- Scalenus triangle is the posterior compartment of costoclavicular space. The division into anterior and posterior compartments is by scalenus anticus. The anterior compartment contains subclavian vein.

KEY BOX 8.5

INTRODUCTION TO THORACIC OUTLET
- Tight space with bones all around
- Brachial plexus and subclavian artery are chief contents
- Scalenus anticus muscle is the muscle for landmark
- Vascular compression is more dangerous
- Neurological symptoms are often undiagnosed

- **Boundaries of scalene triangle** (Fig. 8.2)
  Base : First thoracic rib
  Anteromedially : Scalenus anticus
  Posterolaterally : Scalenus medius
- If the base (the first thoracic rib) is raised by interposition of cervical rib or any other causes (Key Box 8.6), it results in compression of the subclavian artery.

PATHOPHYSIOLOGY OF CERVICAL RIB WITH COMPRESSION

- Due to slow compression, artery distal to the compression dilates due to jet-like effect and turbulence of blood flow. This is described as **poststenotic dilatation** (Venturi effect) (Fig. 8.3 and Key Box 8.7).

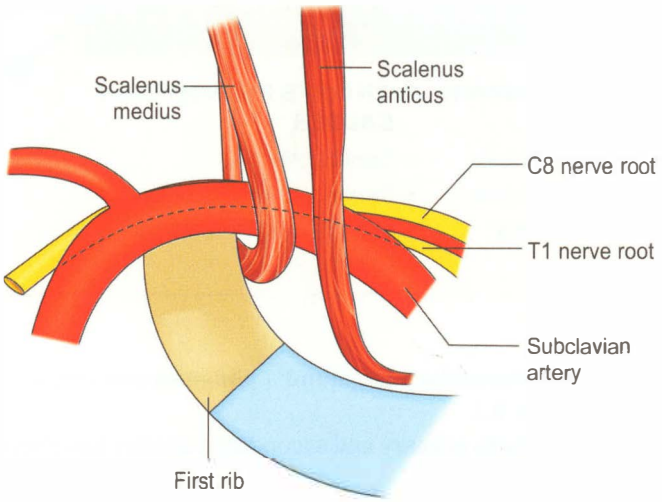


Fig. 8.2: Scalene triangle

KEY BOX 8.6

THORACIC OUTLET SYNDROME—CAUSES
- **T**ransverse process of C7—long
- **H**yperabduction syndrome—compression by pectoralis minor
- **O**perative scars—fibrous bands
- **R**ib—cervical rib
- **A**nomalous first rib—abnormal
- **C**ostoclavicular syndrome—compression between clavicle and first rib
- **I**nsertion of scalenus—anomalous (Scalenus anticus syndrome)
- **C**allus—malaligned fracture clavicle

Remember as **THORACIC**

KEY BOX 8.7

SUBCLAVIAN ARTERY OCCLUSION EFFECTS

Lumen narrowing
↓
Fibrosis or thickening of arterial wall
↓
Stenosis
↓
Poststenotic dilatation
↓
Multiple thrombi → Embolism → Ischaemia

- In this dilated segment, small multiple thrombi develop, which, when dislodged, result in emboli and distal ischaemia. Vascular symptoms are strictly unilateral.

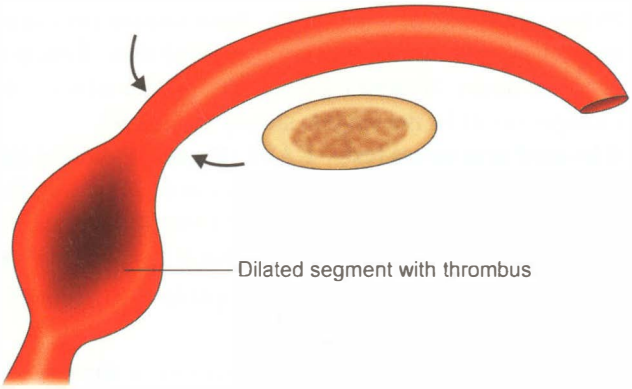


Fig. 8.3: Poststenotic dilatation

Cervical rib

- This is an **extra rib** present in the neck in about 1–2% of the population.
- Commonly unilateral and in some cases it is bilateral.
- It is **more frequently** encountered on the **right side.**
- It is the **anterior tubercle** of the transverse process of the 7th cervical vertebra that attains **excessive development** and results in cervical rib.

Types of cervical rib (Fig. 8.4)

Type I The free end of the cervical rib is **expanded** into a hard, **bony mass** which can be felt in the neck.

Type II **Complete** cervical rib extends from C7 vertebra posteriorly to the manubrium anteriorly.

Type III Incomplete cervical rib, which is partly bony, partly fibrous.

Type IV A complete fibrous band which gives rise to symptoms but cannot be diagnosed by X-ray.

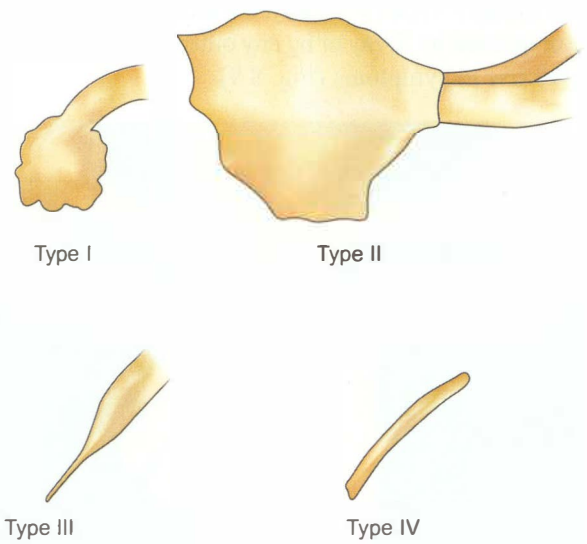


Fig. 8.4: Four types of cervical rib (*see* text)

[1]Card test and Froment's sign are not seen in cases of cervical rib

Clinical features

- Common in **young females**. Even though congenital, symptoms appear only at or after puberty. This is because of the development of shoulder girdle muscles and sagging of the shoulder which narrows the root of the neck. Nerve roots C8, T1 are stretched by completion of growth around 25 years.
- Dull-aching pain in the neck is caused by expanded bony end of cervical rib.
- **Features of upper limb ischaemia**
  - Claudication pain is apparent when the arm with muscle wasting is used. Low temperature, pallor, excessive sweating (vasomotor disturbances), **splinter haemorrhages**, ischaemic ulcers in fingers and gangrene of the skin of the fingers are the other features. Peripheral pulses may be absent/feeble. Oedema and venous distension are very rare. These are called vascular symptoms of cervical rib.
- **Features of ulnar nerve** weakness (lower nerve roots involvement, mainly first thoracic nerve) manifest as tingling and numbness, or paraesthesia in the distribution of C8, T1. The following are the tests which confirm ulnar nerve weakness. It includes sensory disturbances and motor disturbances (performing fine action—writing, buttoning, etc.)
  - **A. Card test[1]:** The patient is asked to hold a thin paper or a card in between the fingers. In cases of ulnar nerve paralysis, due to weakness of interossei muscles, the patient will not be able to hold the card tightly (Fig. 8.5).
  - **B. Froment's sign:** Patient is asked to hold a book between the hand and the thumb. In cases of ulnar nerve paralysis, since the adductor pollicis is paralysed, there is flexion at the distal interphalangeal joint of the thumb.

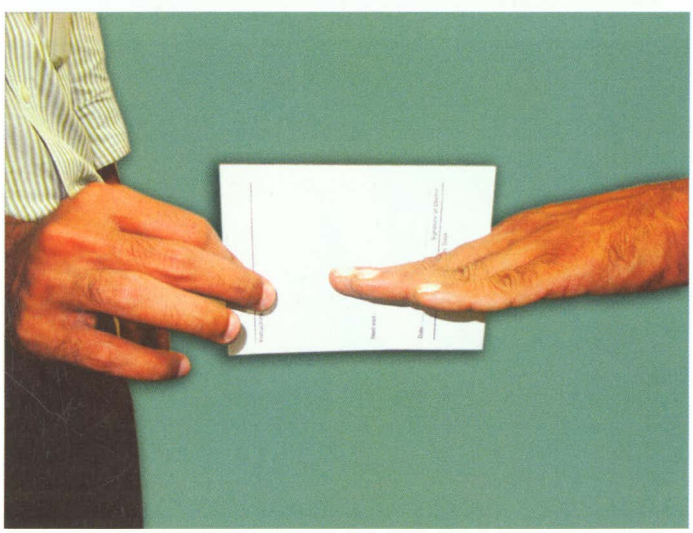


Fig. 8.5: Card test

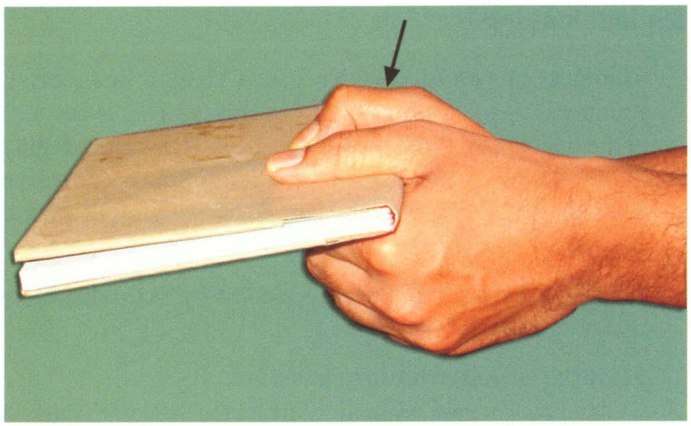


Fig. 8.6: Froment's sign

This is because **flexor pollicis longus**, which is **supplied by median nerve, contracts** (Fig 8.6).

- **Adson's test:** Feel radial pulse, ask the patient to take deep inspiration and turn the neck to the same side. The pulse may disappear or it may become feeble. This test indicates compression on subclavian artery (Fig. 8.7).
- **Hyperabduction test (Halsted test):** This test is done to rule out hyperabduction syndrome caused by pectoralis minor. The radial pulse becomes weak on hyperabduction due to angulation of axillary vessels and brachial plexus, which gets compressed between pectoralis minor and its attachment to the coracoid process.
- **Military attitude test:** When shoulders are set in backward and downward positions the radial pulse becomes weak. This is due to the compression of subclavian artery between the clavicle and the first rib. **This is seen in costoclavicular syndrome.**
- **Allen's test:** Ask the patient to clench his fist tightly and compress the radial and ulnar arteries at the wrist with the thumbs. Wait for 10s and ask the patient to open his hands. Pallor can be seen in the palm. Now release pressure on the radial artery and watch for the blood flow. Repeat test for ulnar artery. If there is occlusion of either artery, colour changes occur in the fingers slowly (Fig. 8.8).
- **Elevated arm stress test—EAST (Roos'):** Patient is asked to abduct the shoulders to 90 degrees and to flex the elbow. Then he is asked to pronate/supinate forearms continuously. Appearance of symptoms suggests thoracic outlet syndrome.
- **A hard mass** may be visible or palpable in the root of the neck (Type I).
- On palpation of supraclavicular region, a **thrill** and on auscultation, a **bruit** can be heard in cases of poststenotic dilatation.

Differential diagnosis

A patient who presents with a few neurological symptoms and signs in the upper limb with a cervical rib may be having some other causes for those symptoms. Hence, it is important to exclude other causes.

1. **Cervical spondylosis:** This should be considered as a possibility in patients above the age of 40 years.
2. **Cervical disc protrusion and spinal cord tumours** may mimic cervical rib with predominant neurological features.
3. **Carpal tunnel syndrome** can occur due to various causes such as myxoedema, rheumatoid arthritis and malunited Colles' fracture. Predominant features of median nerve involvement, more so in menopausal women gives a clue to the diagnosis.
4. **Raynaud's phenomenon**
5. **Costovertebral anomalies**
6. **Pancoast tumour.**

Investigations

1. **X-ray neck** may show a cervical rib (Types I, II and III). Interestingly Type IV variety, a fibrous band which cannot be diagnosed by X-ray or by any other investigation, usually gives rise to symptoms (Fig. 8.9).

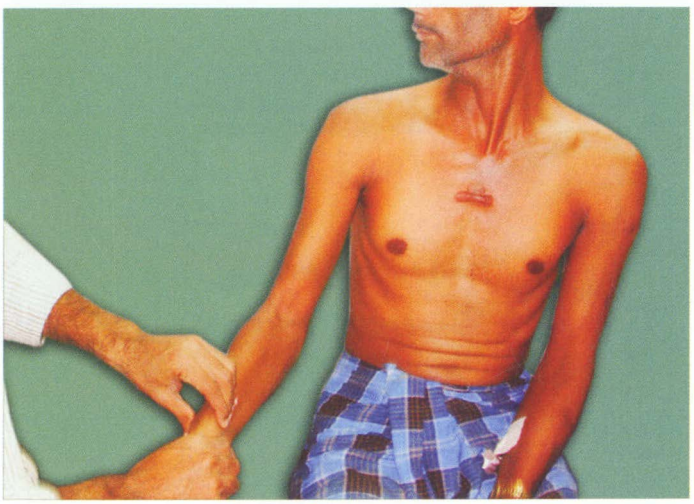


Fig. 8.7: Adson's test

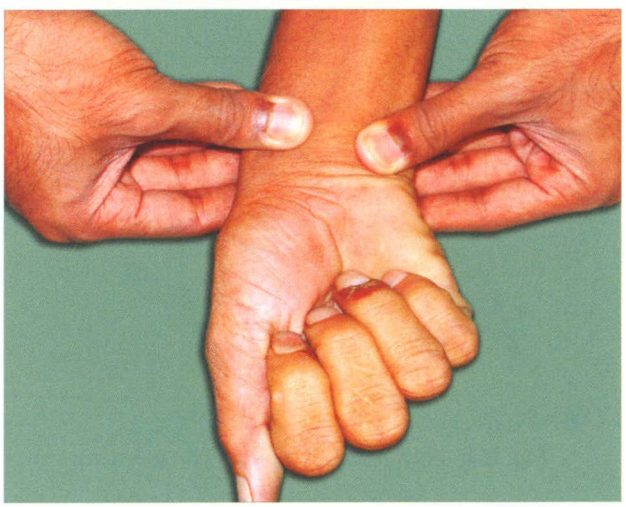


Fig. 8.8: Allen's test

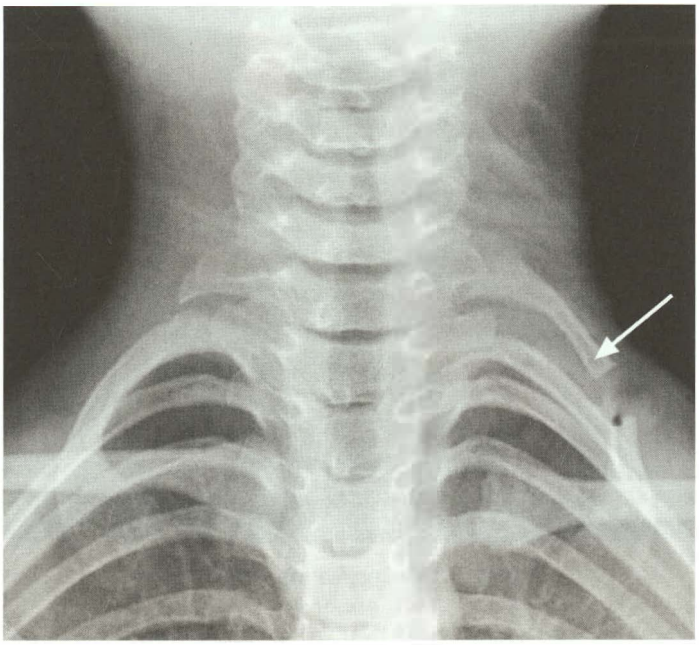


Fig. 8.9: X-ray of the neck showing cervical rib

2. Cervical disc protrusion and spinal cord tumours may have predominant neurological features and thus may mimic cervical rib. **When in doubt, ask for MRI.**
3. **Duplex scan** of affected limb, to detect any aneurysm.

Treatment (Fig. 8.10)

I. Conservative

- Patients with mild neurological symptoms are managed by shoulder girdle exercises or correction of faulty posture.

II. Surgery (Key Box 8.8)

- Presence of **vascular symptoms** and signs are the definite indications for surgery.
- Excision of cervical rib including periosteum is called **extraperiosteal excision** of cervical rib (so that it will not regenerate). This is combined with **cervical sympathectomy** if vascular symptoms are predominant.
- If there is a thrombus in the subclavian artery, it is removed and the artery is repaired (Fig. 8.11).

KEY BOX 8.8

CERVICAL RIB SURGERY (Figs 8.12 and 8.13)

- Remove cervical rib
- Repair subclavian artery
- Restore circulation
- Reduce vasospasm—sympathectomy
- Recognise other causes

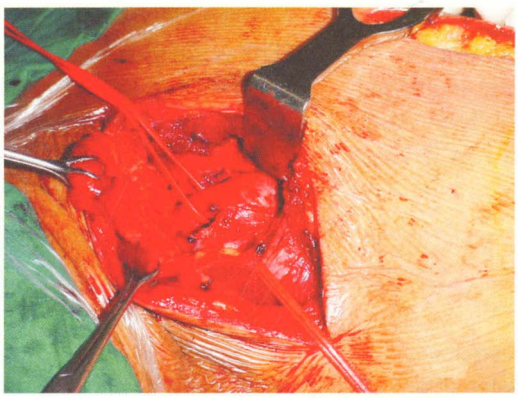


Fig. 8.11: Subclavian artery is exposed to repair the dilatation and remove the thrombus

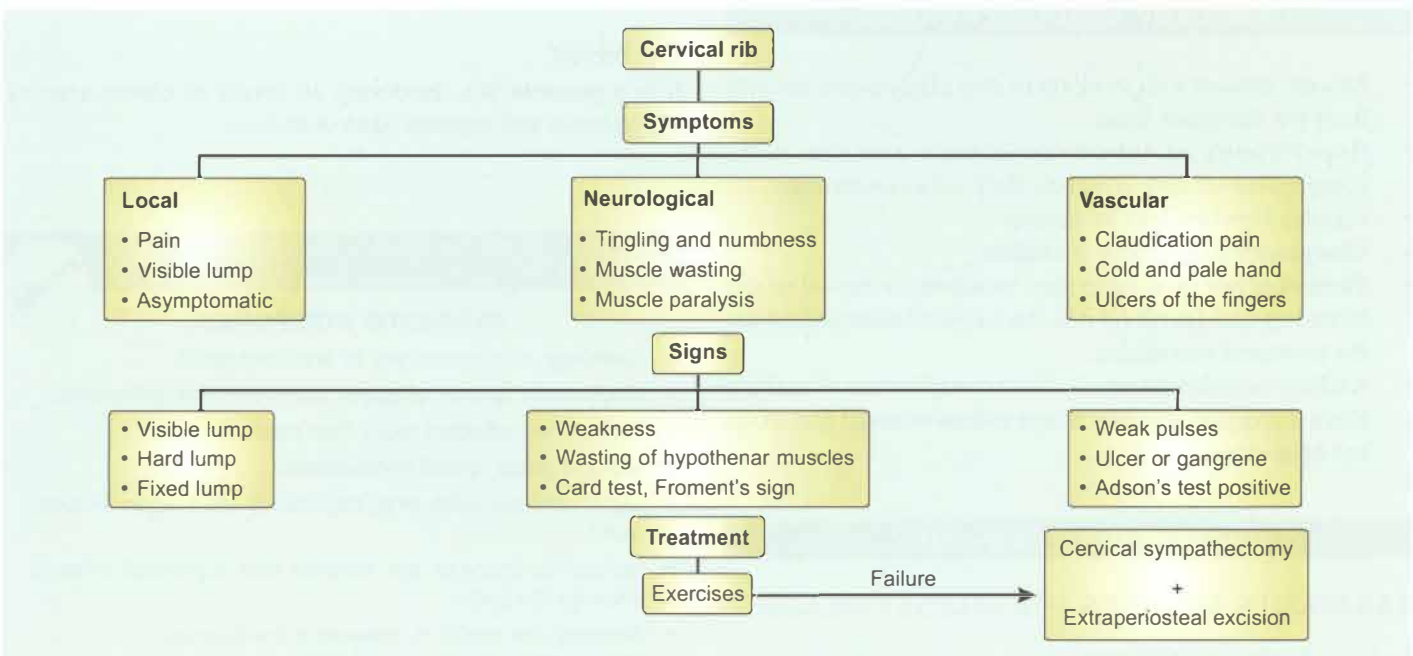


Fig. 8.10: Summary of cervical rib

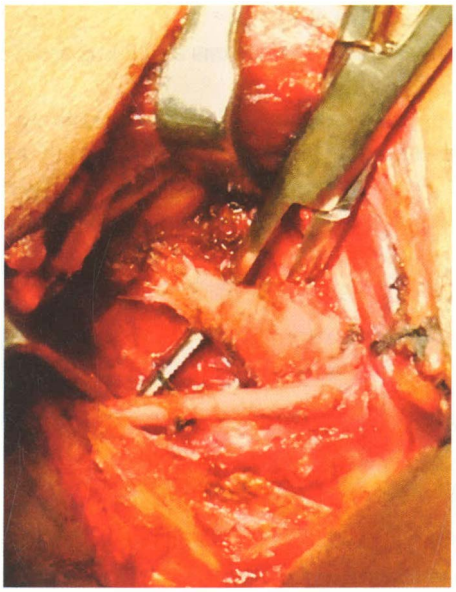


Fig. 8.12: Cervical rib at surgery

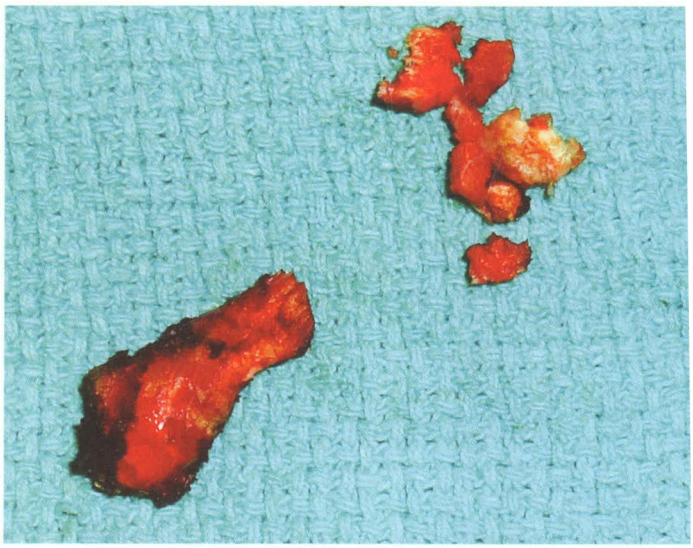


Fig. 8.13: Cervical rib removed

- At exploration, if cervical rib is not found, scalenus anterior muscle is divided. This is called scalenotomy (Key Box 8.9). If **hyperabduction syndrome** is diagnosed, **pectoralis minor is divided** from its insertion into the coracoid process.

KEY BOX 8.9

IF CERVICAL RIB IS NOT FOUND

- Scalenotomy
- Division of pectoralis minor
- Extraperiosteal resection of the first rib

AXILLARY VEIN THROMBOSIS

- Patients present with swelling of arm after intense activity from the dominant hand
- **Hypertrophy of subclavius muscle** also can cause compression of subclavian-axillary vein (sportsman)
- Peripheral pulses will be normal
- Venography to diagnose thrombus
- **Thrombolysis** or if necessary venotomy, removal of the thrombus and 1st rib (if it is the cause of obstruction) are the treatment modalities.
- Axillary vein thrombosis is also a complication of axillary block dissection specially where extensive nodal dissection has been done.

VASCULITIS SYNDROMES

TAKAYASU'S ARTERITIS (PULSELESS DISEASE)

- It is of unknown aetiology.
- Commonly affects females (85%).
- It is a panarteritis involving aortic arch and its branches. Subclavian artery is involved in 85% of the cases.

Clinical features

It starts as a generalised inflammatory disease—fever, body ache, malaise and arthralgia

- Upper limb claudication (Key Box 8.10)
- Absence of peripheral pulses
- Hypertension is common in 50% of the cases due to renal artery involvement.
- Bruit may be heard over the subclavian artery
- Visual disturbances can occur due to involvement of retinal arteries. Late blindness can occur.

Pathology

It is a **panarteritis**, involving all layers of elastic arteries. Thrombosis and stenosis can occur later.

KEY BOX 8.10

VASCULITIS SYNDROMES

- Aetiology is inflammatory or immunological.
- Uncommon causes of upper and lower limb ischaemia.
- Women are affected more than men.
- Multiple small vessel involvement.
- Symptoms are confusing, depending upon organ involvement.
- Ischaemic changes are minimal and superficial when it involves the limbs.
- Steroids are useful in controlling the disease.
- Immunosuppression should be tried carefully.

Investigations

- C-reactive protein is elevated as part of acute phase response (nonspecific).
- **Duplex-Doppler ultrasound, MR angiography** can diagnose the site of obstruction and blood flow pattern.

Treatment

- Very early cases benefit with steroid therapy, e.g. tablet **prednisolone 30 to 50 mg/day** (anti-inflammatory effect). Cyclophosphamide can be tried when other measures fail (immunosuppressive effect).
- Vascular reconstruction—difficult.

GIANT CELL ARTERITIS

- It is also called **temporal arteritis**.
- Elderly women presenting with severe headache is the common presentation.
- Fever and malaise may also be present.
- Involvement of various arteries will result in various symptoms (Key Box 8.11).
- Palpable, pulsatile, tender temporal arteries will clinch the diagnosis.

KEY BOX 8.11

VESSEL INVOLVED	SYMPTOMS
• Temporal artery	Headache
• Facial artery	Jaw pain
• Retinal artery	Sudden blindness
• Upper limb artery	Claudication
• Coronary artery	Myocardial infarction

- Biopsy of the temporal arteries will reveal giant cell granuloma, comprising mainly CD4+ T lymphocytes.
- Treated with prednisolone 60–80 mg/day and slowly tapered over 1–2 years.
- Relapses and remissions are common.

POLYARTERITIS SYNDROME

- This includes microscopic polyarteritis (commonly) and polyarteritis nodosa (less often).
- This syndrome also has an inflammatory reaction.
- Ischaemia of the lower limbs and upper limbs can occur due to involvement of small vessels.

KEY BOX 8.12

PREGANGRENE—SYMPTOMS

- Rest pain is the main symptom
- Pallor on elevation
- Congestion on dependent position
- Guttering of veins
- Poor capillary filling
- Thickening or scaling of skin

- Abdominal pain is due to involvement of visceral vessels.
- Involvement of renal arteries causes loin pain, haematuria and hypertension.
- Treatment is similar to other diseases mentioned above.

SYSTEMIC SCLEROSIS—SCLERODERMA

- Earlier called collagen vascular disorder because of obstruction of the small vessels by collagen deposition.
- Now included under vasculitis syndromes because of their association with inflammatory reaction.
- Ischaemic changes occur in the fingers and toes. Necrosis and ulceration are common.
- Oesophageal involvement results in dysphagia.
- Small bowel sclerosis results in disordered motility and malabsorption.
- Sympathectomy and vasodilators may be useful.
- Raynaud's symptoms can be controlled using calcium channel blockers and nitrates.

GANGRENE

Definition

Macroscopic death of tissue with superadded putrefaction. It affects the limbs, intestines, appendix, etc. In this chapter differential diagnosis of causes of gangrene of the limbs is considered (Figs 8.14 to 8.19).

Pregangrene (Key Box 8.12)

Rest pain, colour changes at rest and with exercise, oedema, hyperaesthesia, skin ulcerations are due to inadequate blood supply to the limb. These changes are described as pregangrenous changes in the limb.

Classification of gangrene

1. **Cardiovascular causes**
   - TAO
   - Atherosclerotic gangrene
   - Acute embolic gangrene
   - Syphilitic gangrene
   - Raynaud's disease
   - Cervical rib
   - Vasculitis syndrome
   - Polycythemia
2. **Neurological causes:** Hemiplegia, paraplegia, bedsore
3. **Traumatic gangrene**
   - Direct—thrombosis; indirect—crush injuries
4. **Physical causes:** Sunrays, radiation, corrosive acids
5. **Drugs:** Ergotamine
6. **Diabetic** gangrene
7. **Acute infective gangrene:** Boil, carbuncle, cancrum oris, gas gangrene.

VARIOUS TYPES OF GANGRENE (Figs 8.14 to 8.19)

Fig. 8.14: Peripheral gangrene due to sepsis and vasopressors

Fig. 8.17: Left medial 2 fingers gangrene due to TAO

Fig. 8.15: Emboli at the brachial artery resulting in massive gangrene of the hand (wet gangrene)

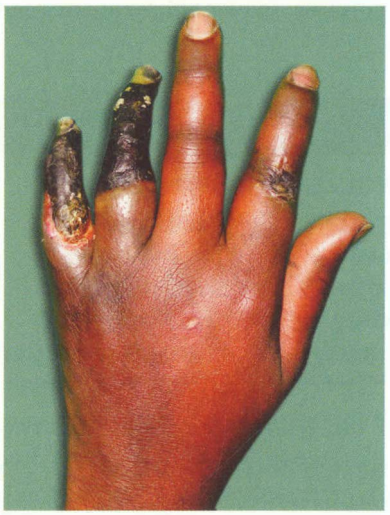


Fig. 8.18: View of the dorsum of the hand

Fig. 8.16: Case of SLE. Observe pulp of fingers

Fig. 8.19: Embolic gangrene of fingers

Clinical features of gangrene

- A part which is gangrenous is a dead portion of the body. It has **no arterial pulsations**, venous return or capillary filling.
- It has no sensation.
- The colour initially will be pale and later it changes to dusky grey and finally black. The **black colour** is due to disintegration of haemoglobin and formation of iron sulphide.
- Signs of gangrene (Key Box 8.13).
- The gangrenous part has to be treated with surgical excision or debridement, which may amount to either disarticulation of the toe or even an amputation.
- The gram-positive, gram-negative and anaerobic organisms multiply in this segment and can produce septicaemia. Thus, this may precipitate multiorgan failure including renal failure, adult respiratory distress syndrome (ARDS), cardiac failure, etc.

KEY BOX 8.13

SIGNS OF GANGRENE

- Loss of pulsation
- Loss of temperature
- Loss of function
- Loss of colour
- Loss of sensation

Clinical types

Basically there are two types—dry gangrene and wet gangrene. They are compared in Table 8.1.

SPECIAL TYPES OF GANGRENE

CANCRUM ORIS

- It is an extensive ulcerative disease of cheek mucosa occurring in malnourished children.
- **Precipitating factors** are:
  - Malnourishment
  - Major diseases such as diphtheria, whooping cough, typhoid, measles and kala azar.
- As a result of these factors the opportunistic organisms such as Vincent's organisms—*Borrelia vincentii* and *B. fusiformis* multiply and cause multiple ulcers, erosions and later, fibrosis.
- Occasionally, as the disease progresses, whole thickness of the cheek may be lost.

Treatment of cancrum oris

1. Ryle's tube feeding
2. Improve nutrition
3. Appropriate antibiotics: Metronidazole 400 mg three times a day for 7–10 days.
4. Reconstructive surgery may be necessary later.

Complications of cancrum oris

1. Fibrosis causing restriction of the movement of jaw.
2. Septicaemia, toxaemia and death.

ACROCYANOSIS

- It is also called hereditary cold extremities.
- Persistent cyanotic discolouration of hands when exposed to cold is a feature.
- This is brought about by intermittent spasm of small peripheral vessels. Commonly affects hands, rarely feet also.
- Generally, it is mild and nonprogressive.

DRUG ABUSE AND GANGRENE

- Abuse of the drugs is an important cause of gangrene in the modern days.
- Inadvertent injection of drugs into artery can lead to thrombosis of the artery resulting in acute ischaemia—commonly in the brachial artery.

Table 8.1 Comparison of dry gangrene and wet gangrene

	Dry gangrene	Wet gangrene
Cause	Slow occlusion of the arteries	Sudden occlusion of the arteries
Involvement of part	Small area is gangrenous due to presence of collaterals	Large area is affected due to absence of collaterals
Local findings	Dry, shrivelled and mummified	Wet, turgid, swollen, oedematous
Line of demarcation	Usually present	Absent
Crepitus	Absent	May be present
Odour	Absent	Foul odour due to sulphurated hydrogen produced by putrefactive bacteria
Infection	Not present	Usually present
Diseases	TAO, atherosclerosis	Emboli, ligatures, crush injuries
Treatment	Conservative amputation	Major amputation is necessary

- Emergency treatment in symptomatic cases include heparinisation and infusion of dextran.
- In severe cases, emergency angiography and intra-arterial thrombolysis is considered.

IATROGENIC DRUG-INDUCED GANGRENE

- Inadvertent **intra-arterial injection of thiopentone** into one of the high divisions of brachial artery, (*congenital anomaly*) usually the ulnar, will result in severe burning and blanching of the hand (Figs 8.20 and 8.21).
- If this complication is noticed following steps (measures) have to be taken immediately.

Case of inadvertent intra-arterial injection (few important steps)

- After the injection, initial signs and symptoms occur very fast (within 15–20 seconds). It consists of intense forearm pain and mottling of the skin on his hand.
- Minutes later, discolouration and nail bed pallor became evident. Approximately 3 to 4 hours later, the symptoms had progressed to paraesthesias and pronounced hand weakness.
- Rapid development of signs indicative of necrosis (by the eighth day) required the patient to undergo fasciotomies, multiple debridements, and 4 skin grafts for cosmesis.

Step 1: If iatrogenic, maintain the intra-arterial catheter in place—do not remove it

Step 2: Identify the progress of the disease—colour changes, necrosis, gangrene

Step 3: Initiate anticoagulation—diluted heparin intravenous and subcutaneous

Step 4: Institute symptomatic relief and plan for rehabilitation—analgesics, physiotherapy

Step 5: Elevation of extremity, antibiotics

Step 6: Perform specific interventions—angiogram, intra-arterial thrombolysis, vasodilators, prostacyclines, sympathectomy, corticosteroids

Step 7: Aim is to save the limb—last will be amputation

Fig. 8.20: Gangrene following intra-arterial injection

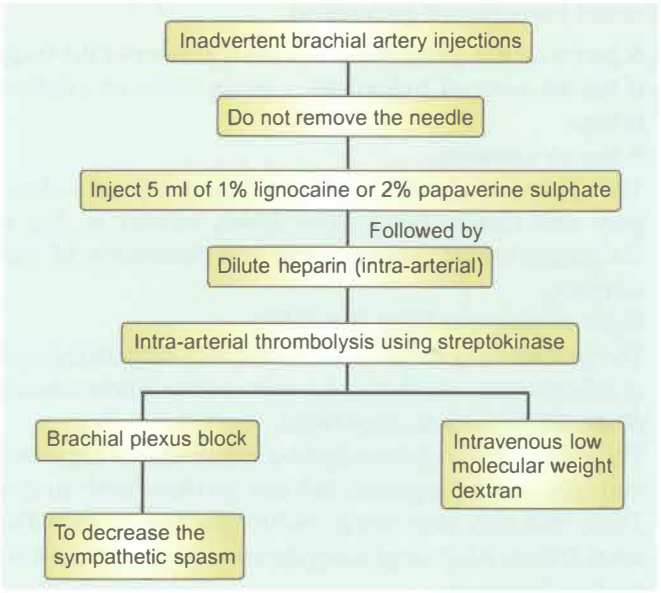


Fig. 8.21: Management of inadvertent brachial artery injections

MISCELLANEOUS

ERGOT AND GANGRENE

- Ergot preparations are used by patients with migraine over a long period.
- Ergotamine gangrene occurs in those who eat bread infected with *Claviceps purpurea.* Example: Dwellers on the shores of the Mediterranean Sea and the **Russian Steppes.**

SUBCLAVIAN STEAL SYNDROME

- Development of symptoms in either brain or in the arm because of subclavian artery obstruction is referred to as subclavian steal syndrome (Key Box 8.14).
- It is most commonly used to describe reversed flow in the vertebral artery ipsilateral to proximal subclavian artery stenosis or occlusion.
- Surgical correction is by endarterectomy or bypass graft.
- Transluminal balloon angioplasty is another alternative.

KEY BOX 8.14

SUBCLAVIAN STEAL SYNDROME

- It is more common on the left side
- **Vertebrobasilar symptoms**—dizziness, vertigo, imbalance
- **Arm symptoms**—fatigue, pain with exercise, paraesthesias, coolness and heaviness on the affected side
- Asymmetrical radial pulses or difference of 20 mmHg systolic pressure between the upper limbs.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- A few Key Boxes and figures have been added.
- Subclavian steal has been discussed.
- A few vasospastic diseases have been added.
- inadvertent intra-arterial injection

MULTIPLE CHOICE QUESTIONS

1. Which of the following is usually not a feature of Raynaud's disease?
A. Radial artery pulsations are absent
B. Upper limb is involved
C. Cervical sympathectomy is helpful
D. Pallor, cyanosis and rubor are the colour changes during the attack

2. Following are true for cervical sympathectomy *except:*
A. It raises the threshold at which spasm occur
B. Entire stellate ganglion has to be removed
C. It is the treatment of choice in hyperhydrosis
D. Up to 3rd thoracic ganglion has to be removed

3. Following muscle is the landmark to thoracic outlet:
A. Scalenus anticus
B. Scalenus medius
C. Scalenus posterior
D. Levator scapulae

4. Following are components of giant cell arteritis *except:*
A. Severe headache is presenting complaint
B. Temporal artery is usually not involved
C. Biopsy from artery reveals giant cell granuloma
D. CD4+ T lymphocytes are found in histopathology

5. Hypertension is a common finding in following conditions *except:*
A. Polyarteritis nodosa
B. Phaeochromocytoma
C. Polycystic disease of kidney
D. Hyperthyroidism

6. Following is the treatment of choice for cervical rib with ischaemia:
A. Excision of the rib
B. Excision of the rib with cervical sympathectomy
C. Excision of the rib with division of pectoralis minor
D. Excision of rib with division of scalenus anticus muscle

7. Following are true for subclavian artery *except:*
A. Cervical rib can compress the artery
B. It gives internal mammary artery
C. Post stenotic dilatation occurs once it is compressed by cervical rib
D. It continues as axillary artery at the medial border of first rib

8. Compression of the subclavian artery is detected by following clinical test:
A. Froment's sign B. Adson's test
C. Allen's test D. Halsted test

9. Following are causes of splinter haemorrhages *except:*
A. Bacterial endocarditis
B. Cervical rib
C. Scleroderma
D. Aortic regurgitation

10. Following are true about subclavian steal syndrome *except:*
A. More common on the left side
B. Present with vertebrobasilar symptoms
C. Both radial arteries are normal
D. Significant arm symptoms are present

11. Following are the causes of upper limb ischaemia *except:*
A. Ergotamine alkaloids
B. Cervical rib
C. TAO
D. Atheroma of the brachial artery

12. Which is the test to detect the dominant arterial supply in the hand circulation?
A. Froment's sign
B. Adson's test
C. Allen's test
D. Halsted test

ANSWERS

1 A	2 B	3 A	4 B	5 D	6 B	7 D	8 B	9 D	10 C	11 D	12 C

9

Lymphatics, Lymph Vessels and Lymphoma

- Lymphoedema—anatomy and physiology
- Lymphatic circulation
- Primary lymphoedema
- Secondary lymphoedema
- Lymphangiography
- Hodgkin's lymphoma
- Non-Hodgkin's lymphoma
- Burkitt's lymphoma
- Sezary's syndrome
- Chyluria
- Immunohistochemistry
- Bone marrow and peripheral blood stem cell transplants
- What is new?/Recent advances

Introduction

Lymphatics and lymph vessels play the role of draining the waste fluid from the body. Hence, they are vulnerable for various infections. The lymphatics are connected to a group of lymph nodes and then drain into the veins. Hence, infections of the lymphatics give rise to enlargement of lymph nodes.

In this chapter, significant surgical diseases affecting the lymphatics and lymph nodes are discussed.

LYMPHOEDEMA

Definition

Accumulation of lymph in the ***extracellular, extravascular compartment*** and subcutaneous tissues results in enlargement of the limb. It is **protein-rich** interstitial fluid.

Common sites of lymphoedema

- Lower limbs are the most common sites.
- Upper limbs
- Scrotum: Elephantiasis of the scrotum is caused by filarial organism *(Wuchereria bancrofti)*.
- Elephantiasis of penis caused by filarial organisms produces ***Ram's horn penis*** (Fig. 9.1).

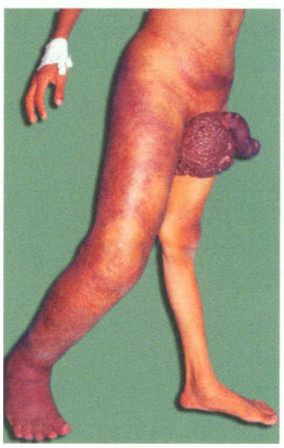


Fig. 9.1: Complication of filariasis—elephant leg, elephantiasis of scrotum and Ram's horn penis

Anatomy and physiology

Functions of the lymphatic system

- To return protein-rich fluid to circulation through ***lymphaticovenous junctions*** in the jugular area.
- This fluid includes water, electrolytes, low molecular weight substances such as polypeptides, growth factors and cytokines.
- It also includes **macromolecules such as fibrinogen, albumin** and globulin.
- **Transport of cholesterol**, long chain fatty acids, triglycerides and fat soluble vitamins (A, D, E, K) by intestinal lymph into circulation. They bypass liver and enter circulation through cisterna chyli and thoracic duct into the left internal jugular vein (lymph from lower limbs, abdomen and left arm).
- Right lymphatic duct drains into right internal jugular vein (lymph from head and right arm).

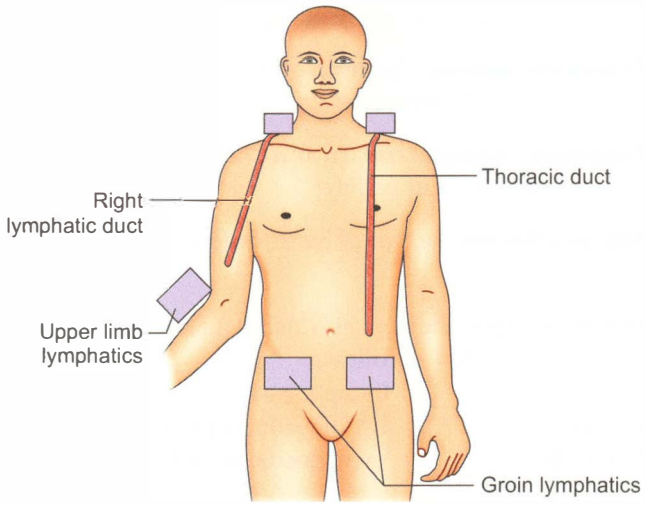


Fig. 9.2: Major lymphatics and lymphatic ducts

Components of lymphatic system

- Lymphatic channels (Fig. 9.2)
- Lymphoid organs—lymph nodes, spleen, Peyer's patches, thymus, tonsils
- Circulating cells—lymphocytes and mononuclear immune cells.

Lymphatic circulation

- Lymph flow is largely due to **intrinsic lymphatic contractility** by lymphangions (segment of lymphatics).
- Transient increase in interstitial pressure secondary to exercise and limb movements also helps to a certain extent.
- Valves prevent reflux in the lymphatics (Fig. 9.3).

Types of lymphoedema

1. Primary lymphoedema
2. Secondary lymphoedema

Risk factors of lymphoedema (Key Box 9.1)

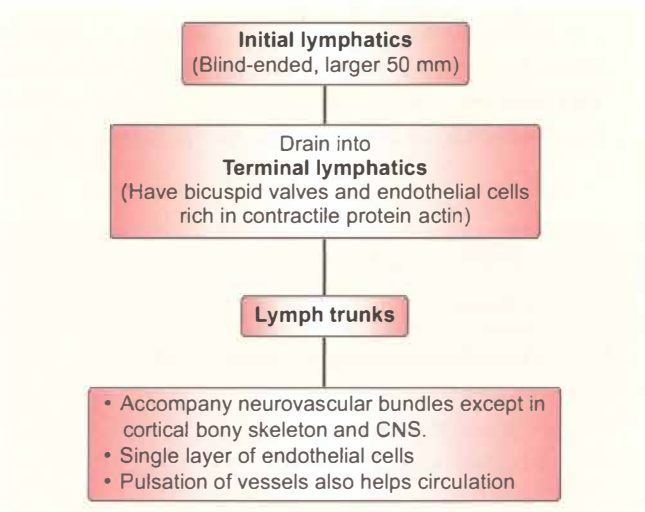


Fig. 9.3: Lymphatic circulation

KEY BOX 9.1

RISK FACTORS FOR LOWER LIMB LYMPHOEDEMA

- Inguinal block dissection, e.g. carcinoma penis, melanoma
- Postoperative pelvic radiotherapy
- Varicose vein stripping and vein harvesting
- Obesity

RISK FACTORS FOR UPPER LIMB LYMPHOEDEMA

- Axillary block dissection, e.g. carcinoma breast
- Radiation fibrosis, scar formation
- Advanced cancer—breast with axillary lymph nodes
- Obesity
- Chronic infection

PRIMARY LYMPHOEDEMA (CONGENITAL)

Introduction

- Incidence is 1 in 6000 persons.
- Lymphoedema is confined to epifascial plane.
- They are due to inherited abnormality of lymphatic system.
- A few cases which occur later in life may be due to unnoticed factors—repeated bacterial and fungal infections, trauma to feet, etc.
- **Loss of venoarteriolar reflux (VAR)** which protects lower limb capillaries from excessive hydrostatic forces in the erect posture, occurs in advancing age. Diseases such as diabetes and chronic venous insufficiency may also contribute.

Causes of primary lymphoedema

I. Hereditary: Associated with syndromes such as Turner's (XO), Klinefelter's (XXY), Down's (Trisomy 21).

II. Familial

1. Nonne-Milroy—type I

- Occurs in 1:6000 live births
- Inherited as autosomal dominant trait
- Brawny lymphoedema of both legs—genitalia, arms, face, etc. develops from birth (Fig. 9.4).

2. Meige's disease—type II

- Lymphoedema develops between puberty and middle age (50 years). It may involve arms also.
- A few are inherited in an autosomal dominant manner.

Pathophysiology (Fig. 9.5)

PEARLS OF WISDOM

Lymphoedema leads to an impairment of immune surveillance and predisposes to other malignancies (Key Box 9.2).

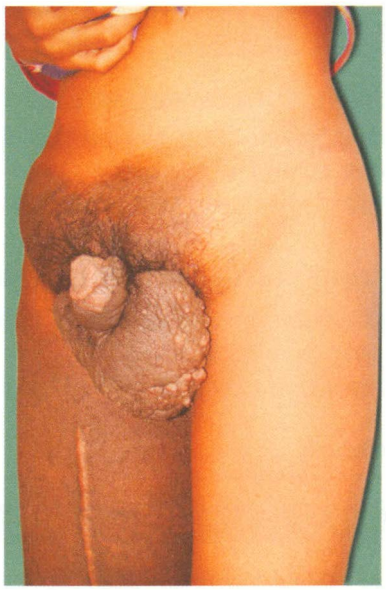


Fig. 9.4: Congenital lymphoedema—observe the right leg. One attempt has been made to decrease lymphoedema by excision

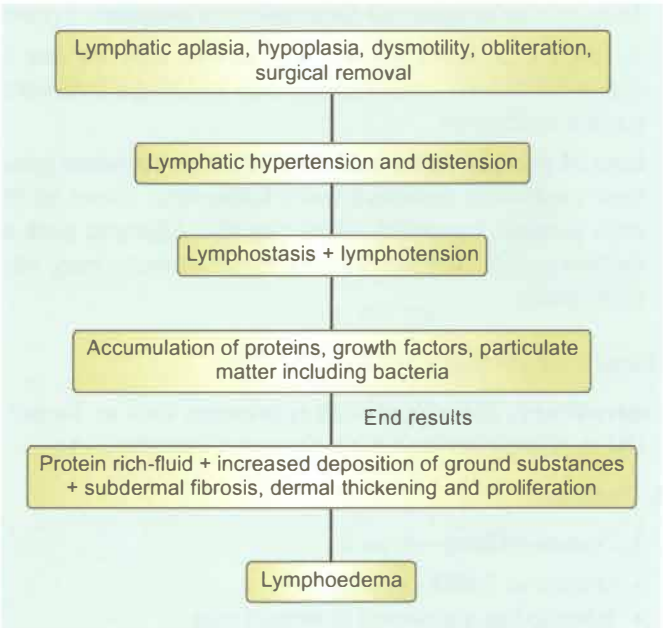


Fig. 9.5: Pathophysiology of lymphoedema

Types of primary lymphoedema

- ***Lymphatic aplasia:*** Number of lymphatic channels and nodes are grossly reduced.
- ***Lymphatic hypoplasia:*** In this variety the lymphatic channels are of small calibre.
- **Milroy's disease** is a type of lymphoedema congenita which runs in families.
- Depending upon the time at which lymphoedema appears it can be classified as follows:

Birth	–	Lymphoedema congenita.
Puberty	–	Lymphoedema praecox.
Later life	–	Lymphoedema tarda.

SECONDARY LYMPHOEDEMA (ACQUIRED)

1. **Filarial elephantiasis** (Fig. 9.6) is caused by *Wuchereria bancrofti,* transmitted by the mosquito *(Culex fatigans).* The disease is caused by adult worms which have the affinity towards lymphatic vessels and lymph nodes. **Microfilariae do not produce any lesion.**
   - Initially, it causes **lymphangitis** which clinically presents with high grade fever, chills and rigors, red streaks in the limb, tenderness and swelling of the spermatic cord and scrotum (Table 9.1).
   - The lymph nodes are swollen and tender. **Retroperitoneal lymphangitis** produces acute abdominal pain.
   - Due to such repeated infections, fibrosis occurs resulting in **lymphatic obstruction**. Later, this gives rise to **lymphatic dilatation**. Lower limb lymphatics are dilated and tortuous (lymphangiectasis).
   - To start with, lymphoedema is **pitting** in nature and progressively becomes nonpitting in nature. Lymph (protein) provides good nourishment for fibroblasts.
   - After repeated infections, the skin over the limb becomes *dry, thickened,* thrown into folds and even *nodules* that *break open* and result in ulcers, Hence, it is called ***'elephant leg'***. Lack of nutrition and infection precipitates lymphoedema. Oedema is also due to reflux of lymph

Table 9.1 Clinical manifestations of lymphatic filariasis

Site of involvement	Acute	Chronic
Lower limbs	Lymphangitis, lymphadenitis	Lymphoedema, chronic lymphadenitis
Scrotum	Lymphangitis	Lymphoedema, chylocoele
Spermatic cord	Acute funiculitis	Chronic thickened cord
Epididymis and testis	Acute epididymo-orchitis	Chronic epididymo-orchitis
Abdomen	Acute retroperitoneal lymphangitis	Chyluria, lymphadenovarix
Breast	Lymphangitis	Lymphoedema

KEY BOX 9.2

MALIGNANCIES ASSOCIATED WITH LYMPHOEDEMA

Skin cancers

- Squamous cell carcinoma
- Malignant melanoma
- Basal cell carcinoma

Sarcomas

- Lymphangiosarcoma (Stewart-Treves syndrome)—sarcoma arising in a lymphoedema after treatment of carcinoma breast
- Kaposi's sarcoma
- Liposarcoma
- Malignant fibrous histiocytoma

Systemic disease

- Lymphoma

Fig. 9.6: Filarial lymphoedema

from para-aortic vessels into the smaller lymphatics draining the lower limb. ***Subcutaneous tissue is grossly thickened. The presence of deep fascia prevents involvement of the deep muscles of the lower limb*** (Key Box 9.3).

2. After inguinal block dissection for secondaries in the lymph nodes (upper limb lymphoedema following axillary block dissection).
3. Following radiotherapy to lymph nodes
4. Advanced malignancies
5. Repeated infections due to barefoot walking (Fig. 9.7).

KEY BOX 9.3

ELEPHANT LEG

Filarial lymphangitis
↓
Lymphatic obstruction
↓
Lymph stasis
↓
Recurrent lymphangitis
↓
Transudation of albumin
↓
Lymphoedema (pitting)
↓
Coagulation of fluid
↓
Repeated infection
↓
Lymphoedema (nonpitting)

Symptoms

- Swollen limb, dull-aching pain
- Tiredness, pins and needles, cramping pain
- Loss of ankle contour
- Buffalo hump on the dorsum of the foot
- Toes are square (Table 9.2)
- Skin on the dorsum of the toes cannot be pinched because of subcutaneous fibrosis—***Stemmer's sign***
- In early cases, it pits on pressure. Later, oedema does not pit and does not reduce even with elevation.
- ***Advanced cases:*** Chronic eczema, fungal infections (dermatophytosis) and nails (onychomycosis), fissuring and warts.
- Ulceration is unusual.
- However, once ulcers start, recurrent bacterial infections are common, thus worsening the disease process.

Table 9.2 Clinical features of lymphoedema

Early signs	Change into	Late/advanced cases—signs
1. Loss of ankle contour	• Fibrosis	1. Chronic eczema
2. Buffalo hump on the dorsum of feet	• Dermal thickening	2. Fungal infection—dermatophytosis
3. **Stemmer's sign:** Skin on dorsum of 2nd toe cannot be pinched because of subcutaneous fibrosis.	• Hyperkeratosis	3. Fungal infection of nails—onychomycosis
4. Early pitting on pressure, less on rest, elevation		4. Fissuring, verrucae, papilla
		5. Grossly thickened leg with skin rugosity—elephant leg.

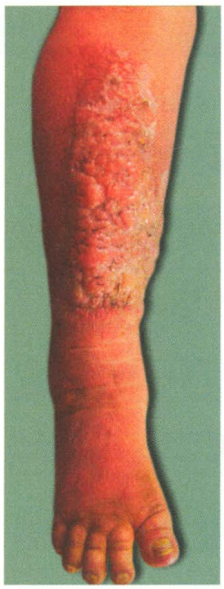


Fig. 9.7: Severe lymphangitis

Grades of filarial lymphoedema

Grade I – **Oedema-pitting:** Completely relieved on rest and elevation. No skin changes.

Grade II – **Oedema-pitting:** Partially relieved on rest and elevation. No skin changes.

Grade III – **Oedema-nonpitting:** Skin involvement, subcutaneous thickening present.

Grade IV – **Oedema-nonpitting:** Not relieved, warty projections, elephantiasis, lymphorrhoea present.

Differential diagnosis of unilateral elephantiasis of the leg

1. **Filariasis** is the most common cause of elephantiasis of the leg in endemic areas such as coastal Karnataka, coastal Andhra Pradesh, Tamil Nadu, etc. (Table 9.3).
2. **Congenital A-V fistula** can present with unilateral gigantism of the leg. Dilated veins, continuous murmur, gigantism, nonhealing ulcer in the leg in a young boy give the clue to the diagnosis.
3. **Elephantiasis neuromatosa** of the leg can cause diffuse enlargement of the leg. The leg is tender on palpation with soft to firm diffuse swelling.
4. **Extensive lipomatosis** of the leg.

Investigations (Lymphangiography)

By and large, no investigations are done in secondary lymphoedema. In selected cases of primary lymphoedema, investigations can be done but are largely of academic interest.

Comparison of primary and secondary lymphoedema is shown in Table 9.3.

Causes of lymphoedema are shown in Key Box 9.4 and Table 9.4.

KEY BOX 9.4

SUMMARY OF CAUSES OF LYMPHOEDEMA

- **A**plasia, hypoplasia (familial and genetic)
- **P**arasitic (filariasis)
- **L**ymph node obstruction—advanced malignancies
- **A**ltered motility—dysmotility (genetic)
- **S**urgical extirpation—block dissection
- **I**nflammatory/Infection—recurrent
- **A**fter radiotherapy, after barefoot walking 'silica particles'.

Remember as **APLASIA**

LYMPHANGIOGRAPHY

Lymphangiography is an investigation wherein a dye is injected into the lymphatics and the entire draining lymphatics and lymph nodes are visualised.

Indications for lymphangiography

1. Lymphoedema, if surgery is planned.
2. In cases of lymphoma, to detect pelvic nodes, para-aortic nodes, etc. but **now CT scan is preferable.**

Procedure

- Commonly, pedal lymphangiograms are done.

Table 9.3 Comparison of primary and secondary lymphoedema

Primary lymphoedema	Secondary lymphoedema
It is due to congenital aplasia and hypoplasia	Filariasis is the common cause
Slowly progressive	Rapidly progressive
It is seen in younger age group	Middle age group
Females are more often affected	Males are more commonly affected
Unilateral, begins distally, spreads proximally	Sometimes, it can start proximally—unilateral or bilateral
Capillary haemangioma may be present	Absent
Regional lymph nodes are absent	Lymph nodes are grossly enlarged
Excisional operations are indicated	Excisional operations and other types of surgery

Table 9.4 Causes of secondary lymphoedema (Figs 9.8 to 9.10)

	Cause	Pathology
1. Infections	Filariasis Tuberculosis Chronic lymphadenitis	Recurrent lymphangitis Lymphadenitis Lymphatic destruction
2. Surgery	Lymph node block dissections (inguinal or axillary)	Excision of lymph nodes as part of treatment
3. Lymph node obstruction	Postradiotherapy large nodes—metastasis lymphoma	Scarred and obliterated lymphatics
4. Tissue damage	Burns scarring	Loss of lymphatics
5. Venous diseases	Chronic venous insufficiency, venous ulcer	Lymphatic destruction
6. Endocrine	Pretibial myxoedema	Obliteration of initial lymphatics by mucin

Fig. 9.8: A case of carcinoma of the breast presented with lymphangitis

Fig. 9.9: Postmastectomy lymphoedema of the right upper limb

Fig. 9.10: Postmastectomy lymphoedema—progressive since 6 years

- 5–10 ml of methylene blue (patent blue) is injected into the web spaces intradermally between the toes. This delineates the lymphatics of the dorsum of the foot which are identified. Then, an oily dye such as "ultra fluid lipoidol" is injected (10–15 ml).
- It may take 12–24 hours to delineate inguinal nodes and para-aortic nodes.
- Isotope lymphangiography refers to injection of albumin labelled with technetium 99m colloid or ^{131}I.

Results

- Metastases appear as irregular filling defects in the lymph nodes.
- It may demonstrate hypoplasia or hyperplasia as in primary lymphoedema.
- If there is obstruction, the dye may return back (dermal back flow).

Lymphangiographic classification

Three types are recognised

1. **Congenital hyperplasia (10%):** This is the condition wherein lymphatics are increased in number. These ***megalymphatics*** are defective resulting in chylous ascites, chylothorax and end in intestines resulting in protein loss (protein-losing enteropathy). Being congenital, it manifests at birth, is progressive, the whole leg is involved and shows variable response to compressive therapy.
2. **Distal obliteration (80%)** (Key Box 9.5)
3. **Proximal obliteration (10%)**
   - It is less common
   - Obstruction is at aortoiliac or inguinal nodes region.
   - Whole leg/thigh is involved (Fig. 9.11).
   - No family history
   - Rapid progression and poor response.

Complications of lymphangiography

1. Lymphangitis and toxaemia
   - It is not being done routinely nowadays, because of availability of ultrasound, CT scan and other noninvasive investigations.

KEY BOX 9.5

DISTAL OBLITERATION (80%)

Distal lymphatics are commonly affected
In and around puberty—praecox involved
Sex: Common in females
Treatment is by compression, good response
Ankle, calf region is involved
Lymphatics are decreased or absent (aplasia)
Remember as **DISTAL**

Fig. 9.11: Unilateral limb oedema due to proximal obliteration

2. Rapid infusion is to be avoided for the fear of lipid pulmonary embolus.

Lymphoscintigraphy

- It has a sensitivity and specificity of 92% and 100% respectively.
- It has replaced lymphangiography.
- Radiolabelled (technetium 99m) colloid is injected into web space between 2nd and 3rd toes or fingers. Limb is exercised periodically and images are taken.
- If there is abnormal accumulation of tracer with collaterals, it is a sign of lymphoedema.
- MRI and CT scan are the latest investigations in addition to lymphangiography for the evaluation of gross swelling of the limb.

Treatment of lymphoedema

PEARLS OF WISDOM

Surgery has a small role in lymphoedema

I. Conservative

1. **Control of swelling:** Decongestive lymphoedema therapy (DLT)
   - Bedrest, elevation
   - ***Bandaging:*** Multilayered lymphoedema bandaging (MLLB—Key Box 9.6).
   - Compression garments

KEY BOX 9.6

MLLB

- Graduated pressure highest at ankle (100%) to lowest in the groin (40%) should be applied.
- It requires about 40–60 mmHg to leg and 30–40 mmHg to arm.
- MLLB should be worn at the start of the day and removed at bed time.
- However, limb should be kept elevated at night.
- The aim is to get a graduated pressure in applying. If it is applied wrongly, it may result in increase in lymphoedema.

   - ***Massage:*** Manual lymphatic drainage (MLD)
   - It aims at draining fluid and protein from ISF space.
   - It also stimulates lymphangion contraction.
2. **Care of skin** (Key Box 9.7)
   - Wash daily with warm water
   - Moisturiser or liquid paraffin should be applied for dry skin
   - Avoid injuries
   - Hyperkeratosis should be treated by keratolytic agent such as 5% salicylic acid.
   - Avoid ointments/herbal medicines/soaps, etc. which are allergic.
   - Antifungal treatment
   - Treat infections early and effectively.
3. **Relief of pain**
   - Pain is multifactorial. It can be due to swelling, infection, involvement of nerve or bone, etc.
   - However, it is often psychosomatic and affected by mood and morale.
   - Antianxiety drugs may help.
4. **Control of infections**
   - Usually it is due to streptococci/staphylococci.
   - Oral or injectable penicillin and flucloxacillin or clindamycin are the drugs of choice.
   - Other drugs are co-amoxiclavulinic acid, cephalosporins.
5. **Exercises**
   - 40% of lymph is formed within skeletal muscle.
   - It is directly proportional to central inflow.
   - Slow systemic, isotonic movements such as swimming and massage will increase venous and lymphatic return.
   - Foot end of the bed should be elevated.
6. **Drugs**
   - **Oxerutins** are the drugs used for venous disease. These are not yet licensed in the UK but are used in India.

KEY BOX 9.7

GUIDELINES FOR TREATMENT OF A PATIENT WITH LYMPHOEDEMA

- **L**
- **Y** Lymphoedema bandage is multilayered
- **M**anual lymphatic drainage (MLD) or massage
- **P**rophylactic antifungal treatment to prevent athlete's foot.
- **H**ygiene of skin and foot
- **A**dvice on diet—weight reduction
- **T**reatment of infection—early and aggressive
- **I**nstructions about exercises
- **C**ompression garments
- **S**urgery consultation as and when required.

Remember as **LYMPHATICS**

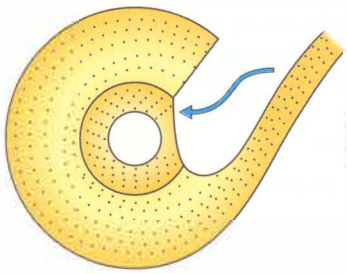


Fig. 9.12: Swiss-roll operation—skin flap is raised

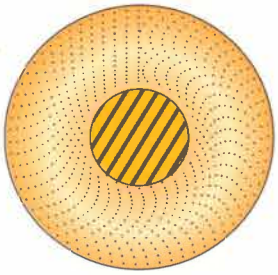
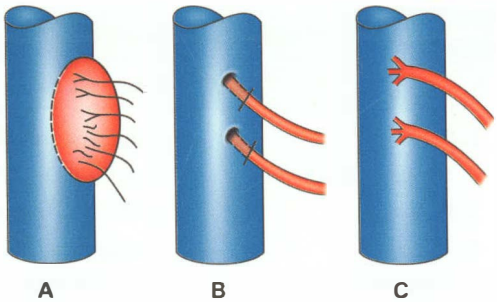


Fig. 9.13: Excision and grafting



Figs 9.14A to C: (A) Node to vein, (B) Lymphatics threaded into a vein and (C) Microvascular anastomosis

- They help by reducing capillary permeability, **improve microcirculatory perfusion**, stimulate interstitial macrophage proteolysis and reduce erythrocyte and platelet aggregation.
- **Diuretics:** 20 mg of furosemide every day/alternate days. This helps in early cases of lymphoedema.
- **Antifilarial treatment:** Diethyl carbamazine citrate (DEC) 100 mg 3 times/day for 21 days with every attack of lymphangitis and once in 6 months.
- **Warfarin** has been used in reducing lymphoedema due to filariasis. It acts by ***enhancing macrophage activity and extralymphatic absorption of interstitial fluid.***
- **Antibiotics** are used in cases of cellulitis and lymphangitis.

II. Surgery

Aim: To reduce the limb size.

1. **Swiss-roll operation (Thompson's):** In this a skin flap is raised containing dermis and it is buried into the deep tissues (close to vascular bundle). This is a dermal flap prepared by denuding epidermis (Fig. 9.12).
2. **Charles excision operation:** It is indicated in primary lymphoedema. It is performed for extensive swelling and skin changes (Fig. 9.13).
   - In this operation, thickened, diseased skin and subcutaneous tissue are excised till the healthy underlying structures are seen followed by split skin grafting.
   - The skin has ***dermal lymphatics which are never involved in filariasis.*** Thus, the subcutaneous lymph may flow *via* dermal lymphatics.
3. **Nodovenous shunt:** Dilated, enlarged lymph node in the inguinal region is anastomosed to a vein nearby, e.g. long saphenous vein or femoral vein (Fig. 9.14).

Thus, these are 3 types of surgery commonly done for filarial leg (Fig. 9.15). There are many other surgeries which are of historical interest. However, the results of surgery for filarial leg are disappointing. Many patients develop intractable ulcers and wetting of the limb due to loss of protein. The wound gets secondarily infected resulting in sepsis, recurrent lymphangitis, etc. As a last resort many patients beg for amputation, to get rid of the 'useless limb.' After amputation the limb can be fitted with prosthesis.

Endemic elephantiasis (Podoconiosis)

- Common in ***tropical*** countries.
- Barefoot walking in the soil during cultivation causes destruction of peripheral lymphatics by ***silica*** particles.

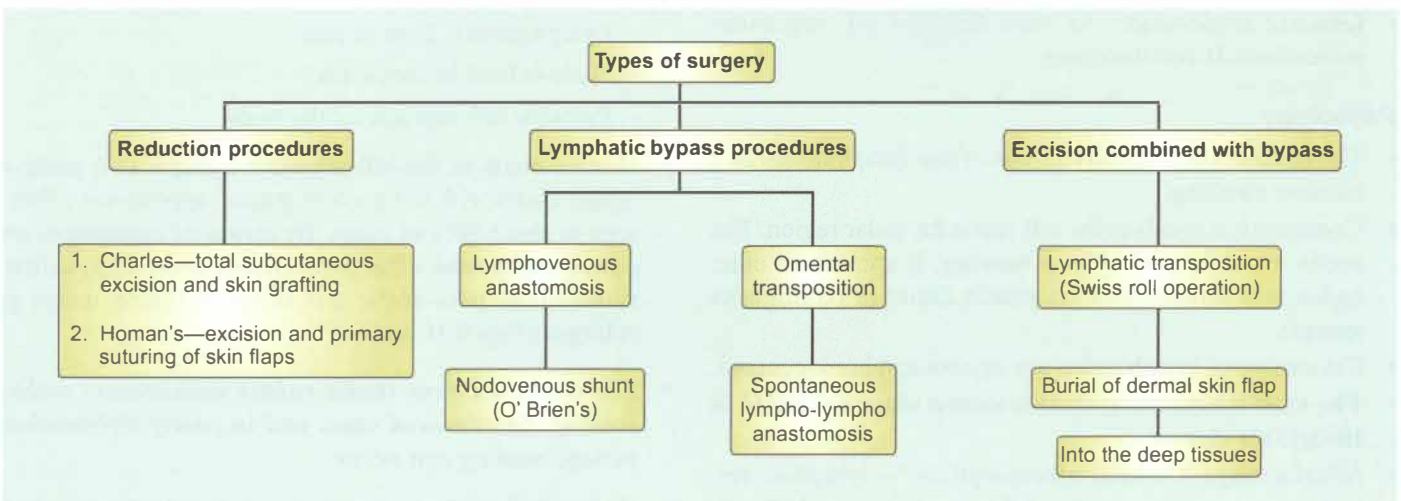


Fig. 9.15: Various surgeries for lymphoedema

Table 9.5 Hodgkin's lymphoma (HL)

	Classical HL	Nodular lymphocytic predominant (HL)
1. Cells	Large atypical cells (Reed-Sternberg cell)	LPHL (popcorn cell—L and H cell)
2. Immunohistochemistry	Express CD15 and CD30 (20 and 45 negative)	Express $CD20^+$, $CD45^+$
3. Progress	Slow (No 'B' symptoms, no bulky disease)	Indolent, favourable prognosis
4. Spread	Contiguous pattern of spread	Does not have contiguous pattern of spread
5. Treatment	Extended field	Involved field RT only
6. Prognosis	Good. Depends on staging	Excellent

- ***Plantar oedema*** develops and spreads rapidly.
- It is a preventable condition and ***wearing of shoes*** can slow down the progression.

Lymphoedema and chronic venous insufficiency (CVI)

- Superficial venous thrombophlebitis (SVT) and deep vein thrombosis (DVT) can lead to lymphatic destruction and secondary lymphoedema. Lymphoedema can predispose to DVT.
- Some degree of superficial reflux may be present in lymphoedema patients that must be managed conservatively rather than by 'blunder surgery'.

HODGKIN'S LYMPHOMA (HL)

Definition

This is a malignant neoplasm of lymphoreticular system. Thus, it can involve lymph nodes, spleen and liver.

Risk factors for Hodgkin's lymphoma

- History of ***infectious mononucleosis*** increases risk of HL two to three fold. ***Epstein-Barr virus (EBV)*** may be a causative agent.
- Slight increase in ***HIV*** cases.
- ***Genetic molecular:*** At least 95% of HL represent monoclonal B cell disorders.

Pathology

- The disease starts usually in one of the lymph nodes as a painless swelling.
- Commonly, it involves the left supraclavicular region. The nodes are ***enlarged without matting***. It spreads to other nodes in a downstream lymphatic drainage (contiguous spread).
- Cut surface of lymph nodes are smooth and homogeneous.
- **The axial lymphatic system is almost always affected in Hodgkin's disease.**
- **Microscopy:** 'Cellular pleomorphism'—lymphocytes, histiocytes, eosinophils and fibrous tissue with **Reed-Sternberg cell,** a giant cell containing mirror image nucleus.

WHO classification/REAL (1994)—Revised European American Lymphoma

I. Lymphocyte predominance, nodular (both Hodgkin's lymphoma and low grade B cell lymphomas)

II. Classic Hodgkin's lymphoma (HL)

- Lymphocyte-rich
- Nodular sclerosis—most common
- Lymphocyte depletion
- Mixed cellularity

However, classical HL and nodular lymphocyte predominant (HL)—LPHL is now more practically used classification (Table 9.5).

Clinical features

- Age: Bimodal distribution. First peak in the 3rd decade and second peak after 50 years.
- Sex: Increased incidence is found in males.
- It presents as generalised lymphadenopathy. Generalised lymphadenopathy means when more than one group of lymph nodes are enlarged and are significant.
- **Significant lymphadenopathy means:**
  - Lymph node > 2 cm in size
  - Node is hard in consistency
  - Palpable left supraclavicular node.
- Disease starts in the left posterior triangle as a group of lymph nodes with a 'bunch of grapes' appearance . This is seen in about 80% of cases. By means of ***contiguous and centripetal spread,*** other lymph nodes in the neck, axillary, mediastinal, para-aortic and inguinal lymph nodes get enlarged (Figs 9.16 and 9.17).
- ***The nodes are firm (India rubber consistency) without matting. In advanced cases and in poorly differentiated variety, matting can occur.***
- Abdominal pain can occur due to hepatosplenomegaly, which are smooth and firm with round borders.

Fig. 9.16: Cervical lymphadenopathy due to Hodgkin's lymphoma. Lymph nodes are firm or rubbery in consistency

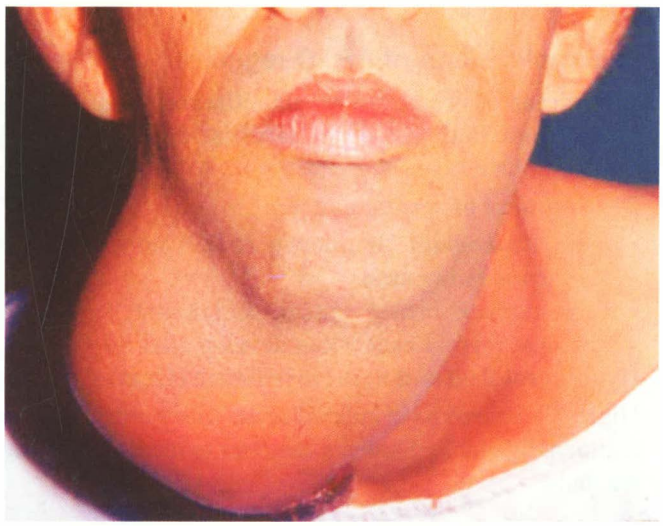


Fig. 9.17: Advanced stage of Hodgkin's lymphoma: Bilateral massive enlargement of nodes

- **Para-aortic nodes** (Key Box 9.8) are felt in the umbilical region, more so on the left side. Its clinical features are:
  - Nodular, firm to hard mass, fixed mass
  - Does not move with respiration
  - Being retroperitoneal, the mass does not fall forward on knee-elbow position.
  - Pulsations may be felt over the mass (transmission from aorta)
- Alcohol-induced pain in involved lymph nodes is a rare symptom.
- There may be ascites.
- **Intermittent fever** (irregular) is sometimes seen. Skin rashes[1] are rare in Hodgkin's lymphoma.
- **Multiple bony pains** can occur due to secondary deposits, especially in the lumbar vertebrae. The secondary deposits are usually osteoblastic giving rise to ***ivory vertebrae***.
- **Superior vena caval obstruction** indicates enlarged mediastinal nodes. This is tested by asking the patient to raise the hand above the head. Engorgement of the veins indicates obstruction and the test is said to be positive ***(Pemberton's test).***

KEY BOX 9.8

PARA-AORTIC NODE ENLARGEMENT COMMON CAUSES

- Lymphoma
- Testicular tumours
- Malignant melanoma
- Gastrointestinal malignancy

Mediastinal lymphoma (Key Box 9.9)

KEY BOX 9.9

MEDIASTINAL LYMPHOMA

- It is the most common malignancy in the mediastinum.
- Anterior compartment is most commonly involved (posterior—rare).
- Most commonly used treatment is chemotherapy/radiation.
- Most dramatic response to the chemotherapy/radiotherapy with a cure rate of 90% in early Hodgkin's and 60% in advanced stage.
- Even in patients with mediastinal bulky lymphoma, symptoms of cough, chest pain, dyspnoea or SVC obstruction is not common.
- **Mediastinal mass ratio (MMR),** defined as the ratio of maximum transverse diameter of mediastinal mass to the maximum transverse intrathoracic diameter, must be calculated. MMR greater than 0.33 by chest X-ray or 0.35 by CT predicts a worse prognosis.

PEARLS OF WISDOM

Generalised pruritus occurs in 25% of patients with Hodgkin's lymphoma. It can be the only presenting feature, preceding the diagnosis by months.

Staging (*see* Table 9.6)

Investigations

1. **Complete blood count (CBC):** Peripheral smear to rule out leukaemia. Anaemia indicates widespread bone narrow metastases.

[1]Mycosis fungoides: It is not a fungal infection but is caused by non-Hodgkin's lymphoma with infiltration of the skin with malignant lymphocytes. Dermatitis and papular rashes are common which progress to tumour formation. It is a variety of cutaneous T cell lymphoma.

Table 9.6 Cotswolds revision of the ann arbor staging system

Stage	Description
I	Involvement of a single lymph node region or lymph node structure
II	Involvement of two or more lymph node regions on the same side of the diaphragm
III	Involvement of lymph node regions or structures on both sides of the diaphragm
IV	Involvement of extranodal sites beyond "E" sites

Annotation description

A—No B symptoms

B—Fever, weight loss > 10% over 6 months, or night sweats

E—Involvement of a single extranodal site contiguous or proximal to known nodal site

X—Bulky disease as defined by > 1/3 widening of mediastinum at T5–T6, or > 10-cm maximum dimension of nodal mass

2. **Elevated creatinine and blood urea nitrogen (BUN)** indicate ureteral obstruction or direct involvement of kidneys (increased uric acid levels indicate aggressive non-Hodgkin's lymphoma).
3. Alkaline phosphatase, calcium and albumin levels. First two are elevated and albumin and haemoglobin levels are lowered.
4. **Chest X-ray** is taken to rule out mediastinal lymph nodes, mediastinal widening, pleural effusion.
5. **Abdominal USG**
   - To look for para-aortic nodes
   - To look for splenomegaly
   - To rule out secondaries in the liver—hepatomegaly
   - However, CT scan of the abdomen is better to define para-aortic nodes when there is minimal enlargement (0.5 cm).
6. **CT scan**
   - CT scan is the investigation of choice for staging.
   - CT scan of chest, abdomen, pelvis is mandatory.
   - CT of the neck is optional.
   - CT will also calculate MMR (*see* to mediastinal lymphoma)
   - CT scan will also help assess renal function and can detect back pressure hydronephrosis.
7. **Lymph node biopsy:** Incision biopsy is done and a neck node is usually removed. Fine needle aspiration cytology (FNAC) may give the diagnosis but a definite histological pattern cannot be made out by FNAC. A trucut biopsy can also give the diagnosis.
8. **Mediastinoscopy** (Chamberlain procedure) is done if peripheral nodes are not available.

Treatment of Hodgkin's lymphoma

1. **Stage I/II classical Hodgkin's:** Low risk
   Chemotherapy + **involved field** radiotherapy
2. **Stage I/II classical Hodgkin's:** High risk

A. ABVD 4 to 6 cycles

– Doxorubicin	25 mg/m^2 IV, days 1 and 5
– Bleomycin	10 mg/m^2 IV, days 1 and 5
– Vinblastine	6 mg/m^2 IV, days 1 and 5
– Dacarbazine	375 mg/m^2 IV, days 1 and 5

Early complications of ABVD regimen

- Adriamycin: Acute cardiac toxicity
- Bleomycin: Pulmonary toxicity
- Vinca alkaloids: Nausea, vomiting, tumour, neutropaenia peripheral neuropathy.
- DTIC: Hair loss

Late complications of ABVD regimen

1. **Life-threatening cardiovascular disease:** This is due to mediastinal radiation—pericarditis, cardiomyopathy cardiac failure.
2. **Development of second cancer**
   - *Acute leukaemia:* Incidence is less now as MOPP regimen is rarely used.
   - *Lung cancer,* breast cancer, melanoma, soft tissue sarcoma, thyroid cancer can develop after 30 years.
3. **Hypothyroidism and hyperthyroidism (Graves' disease)**

High risk patients have bulky disease. There are many other regimens available. Students can refer to medical oncology manuals.

B. MOPP regimen

- **M**echlorethamine 6 mg/m^2 body area on 1st day and 8th day.
- **O**ncovin (vincristine) 1.4 mg/m^2 IV on 1st day and 8th day.
- **P**rocarbazine 100 mg orally, for 1 to 10 days.
- **P**rednisolone 15 mg 8th hourly orally, for 1 to 10 days.
- Minimum of 6 cycles or at least 2 extra cycles after attaining complete remission should be given. In spite of stage III and stage IV disease, survival of 10 years with disease-free interval is about 80% (Key Box 9.10).

Complications of MOPP therapy

- Infertility in both men and women
- Development of acute myeloid leukaemia
- Haematosuppression (bone marrow suppression)

3. **Stage III/IV classical HL**
   - 60 to 70% of patients with advanced stage Hodgkin's lymphoma can be cured with 6 cycles of ABVD chemotherapy which is superior to older MOPP.

KEY BOX 9.10

ADVANCED HL—INDEPENDENT PROGNOSTIC FACTORS

- **A**lbumin less than 4 g/dl
- **L**ymphocytopaenia < 600/mm^3
- **B**lood Hb less than 10.5 g/dl
- **U**nder 44 years of age
- **M**ale sex
- **I**nvolvement of liver, bone (advanced)
- **N**eutrophils (WBC) > 15,000 cells/mm^3, leukocytosis

Remember as **ALBUMIN**

4. LPHL

- Involved field RT alone is for treatment of early stage LPHL.
- Stage III and stage IV are treated by combination chemotherapy.

5. Recurrent Hodgkin's disease

- All patients who are less than 70 years, who relapse after therapy can be considered for autologous **haemato-poietic cell transplant (HCL).**
- It should be remembered that drug therapy and HCL can be offered to those who are fit patients only and toxicity of all the drugs should be explained to these patients.

PEARLS OF WISDOM

Extended mantle field radiotherapy can cause carcinoma of the breast. Pelvic radiotherapy (inverted Y) can cause infertility. Hence, they are not commonly used now.

Complications of disease (natural history)

1. **Mediastinal disease**—pleural effusion, SVC obstruction
2. HL decreases the cell-mediated immunity. It causes depressed CD4+/CD8+ ratio. They are vulnerable for **opportunistic infections**—mycobacteria, herpes zoster, cytomegalovirus.

What is no longer followed in Hodgkin's lymphoma?

1. Staging laparotomy

With availability of CT scan, ultrasound and lympho-scintigraphy, all the details of involvement of intra-abdominal viscera or pathology can be detected. Thus, staging laparotomy is not done any longer.

2. Splenectomy: Spleen is required for immunological functions.

3. Gallium scan: Detection of lymph nodes is by scintigraphy if required.

4. Bipedal lymphangiogram: Same reasons as mentioned above.

5. Intravenous pyelography: Renal function can be easily assessed by CT scan.

6. MOPP regimen: Not favoured—because of toxicity

7. Pelvic RT (responsible for infertility): Not favoured.

8. Classification: Ryle's classification is no longer used.

NON-HODGKIN'S LYMPHOMA (NHL)

Aetiology

1. Age and sex

- Small lymphocytic lymphoma: Elderly patients
- Lymphoblastic lymphoma: Male adolescents and young adults
- Follicular lymphoma: Mid-adult age group
- Burkitt's lymphoma: Children, young adults

2. Viruses

- RNA viruses: Human immunodeficiency virus (HIV) produces AIDS. These patients can develop high grade B cell lymphoma.
- DNA viruses: Epstein Barr viruses (EBV) can produce Burkitt's lymphoma.

3. Bacteria

- *H. pylori:* Gastric extranodal marginal zone B cell lymphomas of MALT type.

4. Immunodeficiency states

- AIDS
- Organ transplantation

Classification of non-Hodgkin's lymphoma

I. Low grade

- Small lymphocytic
- Follicular, predominantly small cleaved cell

II. Intermediate grade

- Follicular, predominantly large cell
- Diffuse, small cleaved cell
- Diffuse mixed small and large cell
- Diffuse large cell

III. High grade

- Large cell immunoblastic
- Lymphoblastic
- Burkitt's or non-Burkitt's lymphoma.

Pathological classification

I. B cell NHL: Small lymphocytic lymphoma, follicular lymphoma, Burkitt's lymphoma (Key Box 9.11).

II. T cell NHL: Cutaneous T cell lymphoma, mycosis fungoides and Sezary syndrome, lymphoblastic lymphoma.

Management of non-Hodgkin's lymphoma

It depends on following factors:

- Grade of tumour
- Stage of disease
  - Stages I and II: Low grade—radiotherapy
  - Stages I and II: Intermediate and high grade—chemotherapy
  - Stages III and IV: Chemotherapy

Investigations

- Complete blood count
- Liver function tests including LDH

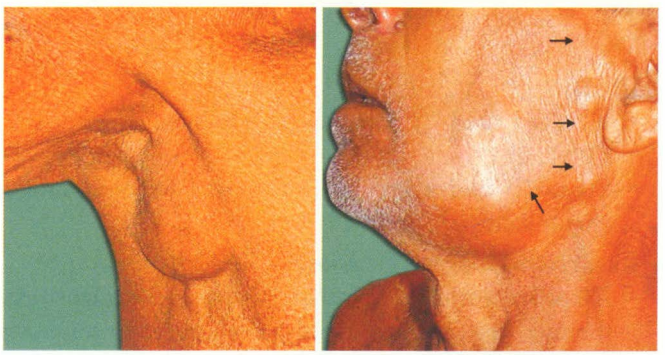
PEARLS OF WISDOM

LDH is an important prognostic factor and aggressive therapies are required for patients with increased LDH.

DIFFERENT SITES OF LYMPH NODES IN NHL (Figs 9.18 to 9.25)

Fig. 9.18: Non-Hodgkin's lymphoma with skin involvement

Fig. 9.19: Large occipital lymph node



Figs 9.20A and B: Massive axillary lymphadenopathy and enlargement of submandibular, upper deep cervical and preauricular lymph nodes

Fig. 9.21: Advanced case of non-Hodgkin's lymphoma with involvement of groin nodes, testicle and skin deposits in the thigh

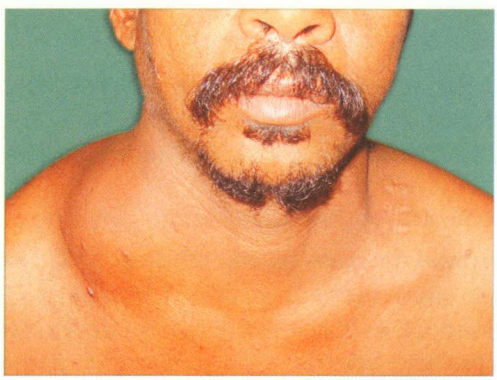


Fig. 9.22: Massive cervical lymph node enlargement. Observe the scar of the lymph node biopsy on the opposite side, reported as normal

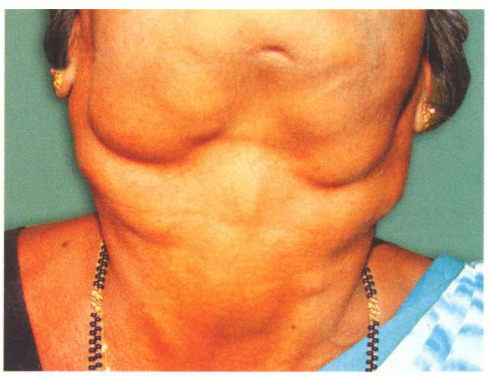


Fig. 9.23: Bilateral lymph nodes in the upper neck

Fig. 9.24: Preauricular, postauricular and facial lymph nodes

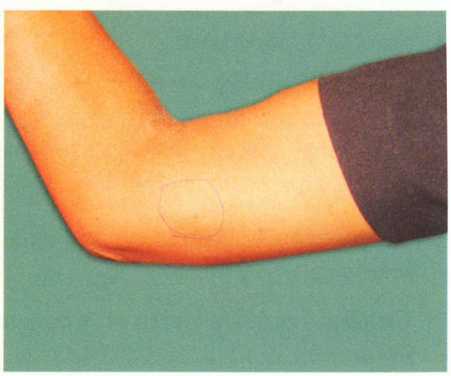


Fig. 9.25: Epitrochlear lymph node enlargement

Table 9.7 Comparison of Hodgkin's and non-Hodgkin's lymphoma

	Hodgkin's lymphoma	Non-Hodgkin's lymphoma
Age	Bimodal age	More than 50 years
Involvement of lymph nodes	Left supraclavicular, axillary, inguinal	External Waldeyer's ring, submental, submandibular, preauricular, postauricular, occipital nodes
Retroperitoneal nodes	Uncommon	Common
Tonsil enlargement	Not enlarged	Tonsils may be enlarged
Epitrochlear node	Not enlarged	Can be enlarged
Hepatomegaly	Uncommon	Common in low grade
Involvement of bone	Uncommon (10%)	In 40% of cases, iliac crest is involved by the time of diagnosis
Alcohol-induced pain	Characteristic	Absent
Fluctuating fever	Present	Absent
Prognosis	Better prognosis	Poor prognosis
Common presentation	Lymphatic organ	Extralymphatic
Mediastinal nodes	Common	Very few patients with DLC (diffuse large B cell lymphoma)

KEY BOX 9.11

FOLLICULAR LYMPHOMAS (FL) OR SMALL LYMPHOCYTIC LYMPHOMA (SLL)

- Called indolent lymphomas
- Can be of many years duration
- Present as painless lymphadenopathy
- Spontaneous regression in 20% of patients
- Also called waxing and waning adenopathy
- 'B' symptoms are unusual.
- Uncommon before the age of 50
- Good prognosis
- Remission duration and survival improvement with **'rituximab'** (anti CD20 antibody specific for B lymphocytes)

- Creatinine, calcium
- Ultrasound, CT scan
- Core needle biopsy, lymph node biopsy and bone marrow biopsy.
- ***Immunohistochemistry*** is required for subclassification. *Examples:* Small lymphatic lymphoma (CD5+, CD23+).
- Mantle cell (CD5+, CD23–), etc.

PEARLS OF WISDOM

If Waldeyer's ring is involved, an upper GI scopy should be considered since the incidence of gastric involvement in such cases is increased.

Treatment

- Combination chemotherapy with COPP (cyclophosphamide, oncovin, procarbazine, prednisolone) or ABVD with or without **rituximab** is the treatment of choice.
- ***Indolent lymphomas*** can also be treated with same regimen with or without involved field RT (IFRT).
- ***Gastric MALT lymphomas*** may completely regress with therapy against *H. pylori* with appropriate antibiotics and proton pump inhibitors.
- ***Comparison of Hodgkin's and non-Hodgkin's*** lymphoma is given in Table 9.7.

MISCELLANEOUS

BURKITT'S LYMPHOMA (SMALL NONCLEAVED LYMPHOMA)

- It is a type of high grade non-Hodgkin's lymphoma first described by Burkitt affecting jaw bone (maxilla, mandible).
- It is rare everywhere except in a few places where malarial infestation is heavy.
- It is caused by ***Epstein-Barr virus*** (EBV) which multiplies in presence of heavy malarial infestation.

Types

- **Endemic:** Parts of Africa and other tropical locations. It is associated with EBV. Affects jaw and orbit. It has good prognosis.
- **Sporadic:** Throughout the world. It is not associated with EBV. It affects abdomen and GI tract rather than bones, has poor prognosis.

Diagnosis

- Biopsy will reveal typical 'starry sky' appearance with primitive lymphoid cells with large clear histiocytes.

Treatment

- Endemic cases respond well to cyclophosphamide.
- Sporadic cases need to be treated with combination chemotherapy.

SEZARY'S SYNDROME

It is also a type of cutaneous T cell lymphomas. Here, skin involvement is manifested clinically as a generalised exfoliative erythroderma along with an associated leukaemia of Sezary cells.

These cells are characterised by cerebriform nucleus. These are indolent tumours having good survival time.

CHYLURIA

- It is the commonest manifestation of rupture of lymph vessels due to filariasis (others being chylocoele, chylous ascites and chylothrax).
- It occurs when intestinal lymphatics are obstructed by filarial fibrosis and the lymphatics are diverted to renal lymphatics.
- The dilated and tortuous lymphatics, due to high pressure, rupture into the renal pelvis and ureter leading to chyluria. Passage of large amount of chyle results in the loss of protein and fat resulting in malnutrition. Such patients are also debilitated with loss of immunity.
- Oral ingestion of fat labelled with Sudan red III turns the urine pink in these cases.
- It is treated by diethyl carbamazine, bed rest, elevation of foot end of bed and administration of high protein diet.

IMMUNOHISTOCHEMISTRY (IHC)

- This is a new technique which detects a specific antigen using an antibody.
- Cytokeratins are expressed by epithelial cells and **cytokeratin positivity suggests carcinoma**.
- Similarly, endothelial cell markers, lymphoid markers, neuroendocrine markers or melanoma markers are available.
- Some **immunohistochemical stains used for tumours** are given below:

  - Epithelial : Cytokeratin
  - Lymphoma : CD3, CD20
  - Melanoma : S100
  - Vascular : CD31, CD34
  - Colorectum : Cytokeratin
  - Ovary : CA125, CK20
  - Prostate : PSA
  - GIST : CD117

- Thus, immunohistochemical stains can detect all types and can confirm malignancy. ***S100 and actin stains*** can be used to identify ***myoepithelial cell layer*** in a duct or gland.
- IHC may also help in selection of treatment and prognosti predictions. *Examples:* Carcinoma of the breast is assesse for oestrogen receptor, progesterone receptor and HER status.
- IHC can also detect infections such as cytomegaloviru (CMV), Epstein-Barr virus (EBV), etc.
- IHC can also be used to detect abnormal accumulation c **proteins** or **amyloid**.

RECENT ADVANCES

Bone marrow and peripheral blood stem cel transplants

- Autologous transplants (which use stem cells from th patient) have the risk of reintroducing lymphoma cells bacl into the patient after treatment.
- New monoclonal antibodies developed for treatin lymphoma may help remove these remaining cells.
- Drugs to eliminate graft-versus-host disease in allogenei (donor) transplants. This work revolves around altering th transplanted T cells so that they will not react with th recipient's normal cells but still kill the lymphoma cells.
- **Targeted therapies: New drugs are**—bortezomit (Velcade), romidepsin (Istodax), and temsirolimus (Torisel)
- **Antibiotics:** Gastric MALT lymphoma, which is linked t infection by the bacteria *Helicobacter pylori*, can often b treated with triple therapy. Doxycycline also is helpful.
- **Monoclonal antibodies:** "Immunotherapy for non-Hodgkin lymphoma." Rituximab is most often given for a short period in follicular lymphoma. Other similar drug such as ibritumomab and tositumomab, epratuzumab, which targets the CD22 antigen on certain lymphoma cells, and obinutuzumab, which targets the CD20 antigen.
- **Immunotoxins:** They act as homing devices to deliver the toxins directly to the cancer cells. One example of this is brentuximab vedotin (Adcetris), which is made up of an antibody to CD30 that is attached to a cell poison. It has been shown to help treat patients with anaplastic large cell lymphoma (ALCL) that is not responding to treatment with chemotherapy.
- **Lymphoma vaccines is used only in clinical trials.**

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- Lymphoedema has been discussed more in length.
- Lymphoma has been updated.
- Immunohistochemistry has been added.
- Recent advances

MULTIPLE CHOICE QUESTIONS

1. Which of the following is not true regarding function of the lymphatic system?

A. Lymph circulation occurs through lymphatico-venous junctions

B. Lymphatic fluid contains macromolecules

C. Right lymphatic duct drains into internal jugular vein

D. Lymphatic fluid from intestines pass through liver

2. Following are true for lymph trunks *except*:

A. It is lined by single layer of endothelial cells

B. Pulsations of the vessels help in lymph circulations

C. Thoracic duct drains into right internal jugular vein

D. It does not accompany neurovascular bundles in central nervous system

3. Following are risk factors for lower limb lymphoedema *except:*

A. Inguinal block dissection

B. Pelvic radiotherapy

C. Obesity

D. Alcohol abuse

4. Stemmer's sign refers to:

A. Gross thickening of the subcutaneous tissue in lymphoedema leg

B. Deep muscle of the leg are not involved

C. Skin on the dorsum of the toes cannot be pinched

D. Buffalo hump on the dorsum of the foot

5. Following is true for filarial elephantiasis of leg *except:*

A. It is caused by *Wuchereria bancrofti*

B. Transmitted by *Culex fatigans*

C. Microfilaria do not produce any lesion

D. Significant changes occur deep to deep fascia

6. Majority of the primary lymphoedema is due to:

A. Congenital hyperplasia of lymphatics

B. Distal obliteration of lymphatics

C. Proximal obliteration of lymphatics

D. Central obliteration of lymphatics

7. Oxerutins used in lymphoedema act by following ways *except:*

A. They decrease capillary permeability

B. Improve microcirculatory perfusion

C. Stimulate interstitial macrophage proteolysis

D. Increase platelet aggregation

8. Which of the following is true in lymphoedema?

A. Dermal lymphatics are never involved in filariasis

B. Oxerutins are not used in lymphoedema

C. Lymphoedema pressure bandage requires 10 mmHg pressure in the leg

D. Lymphoedema does not predispose to malignancies

9. Contiguous and centripetal spread occur in:

A. Hodgkin's lymphoma

B. Non-Hodgkin's lymphoma

C. Burkitt's lymphoma

D. Sezary's syndrome

10. Following are true for lymphocytic predominance nodular lymphoma *except:*

A. It has large atypical cells

B. It expresses CD20+

C. It is indolent

D. It has favourable prognosis

11. Following are true about classical Hodgkin's lymphoma *except:*

A. Spread is contiguous

B. Large atypical cells are seen

C. B symptoms are characteristic

D. Expresses CD15 and CD30

12. Following are the features of Reed-Sternberg cell *except:*

A. It is a giant cell

B. Mirror image nuclei

C. Large multinucleated cell

D. It is not derived from B lymphocytes

13. Following are true for mediastinal lymphoma *except:*

A. It is the most common malignancy in the mediastinum

B. Response to chemotherapy is dramatic

C. It is Hodgkin's lymphoma

D. Posterior compartment is commonly involved

14. Following are no longer followed in lymphomas *except:*

A. Pelvic radiotherapy is given

B. Splenectomy as staging procedure is done

C. Rye's classification is followed

D. ABVD regimen is given commonly

15. Following are causes of epitrochlear enlargement *except:*

A. Hodgkin's lymphoma
B. Non-Hodgkin's lymphoma
C. Secondary syphilis
D. Cat-scratch fever

16. Following are true for Rituximab *except:*

A. It is a monoclonal antibody against protein CD20
B. It is used to treat leukaemia
C. It can also be used to treat follicular lymphomas
D. It has no role in transplant rejections

17. Following are true for Burkitt's lymphoma *except:*

A. It is associated with Epstein-Barr virus
B. It is not associated with heavy malarial infestation
C. Affects jaw bone and orbital bone
D. It gives starry sky appearance in histological sections

ANSWERS

1 D	2 C	3 D	4 C	5 D	6 B	7 D	8 A	9 A	10 A
11 C	12 D	13 D	14 D	15 A	16 D	17 B			

10

Varicose Veins and Deep Vein Thrombosis

- Primary varicose veins
- Secondary varicose veins
- Surgical anatomy of venous system of legs
- Anatomy of the long saphenous vein
- Clinical examination
- Treatment
- Complications
- Short saphenous varicosity
- Deep vein thrombosis
- Recurrent varicose veins
- Pelvic congestion syndrome
- Pulmonary thromboembolism
- What is new?/Recent advances

Introduction

Varicosity is the penalty for verticality against gravity. This is the common statement made in lecture classes. The blood has to flow from the lower limbs into the heart against gravity because of the upright posture of human beings. In many cases, varicose veins are asymptomatic. Raised intra-abdominal pressure also precipitates varicose veins, more commonly in females due to repeated pregnancy. The complications of varicose veins are responsible for hospitalisation of the patient.

Definition

Dilated, tortuous and elongated superficial veins of the limb are called varicose veins.

Examples of varicosity

- Long saphenous varicosity
- Short saphenous varicosity
- Oesophageal varices and fundal varices
- Haemorrhoids
- Varicocoele
- Vulval varix and ovarian varix

In this chapter, varicosity of the leg is discussed.

PRIMARY VARICOSE VEINS

- They occur as a result of ***congenital weakness*** in the vein wall due to defective connective tissue and smooth muscle.
- It can also be due to muscular weakness or due to ***congenital absence of valves*** (Key Box 10.1).
- Very often, the valve at the saphenofemoral (SF) junction is incompetent/absent. The valves can also be absent where the superficial veins join the deep veins.
- **Klippel-Trénaunay syndrome** is a congenital venous abnormality wherein superficial and deep veins do not have any valves. It is also called **valveless syndrome** (Key Box 10.2).
- Primary varicosity can also be **genetic**. Some patients inherit abnormalities in the FOXC2 gene. These factors, in addition to prolonged standing (agriculturists, traffic police, hotel workers), contribute to the development of varicose veins.

KEY BOX 10.1

FACTORS WHICH PREDISPOSE TO VARICOSE VEINS

- **Height:** Tall individuals suffer more
- **Weight:** Obesity may weaken vein wall
- **Occupation:** Hotel workers, policemen, shopkeepers, tailors
- **Side:** Left is affected more than the right
- **Age and sex:** Not very clear

KEY BOX 10.2

KLIPPEL-TRÉNAUNAY SYNDROME

- Incidence 1 in 1,00,000 population.
- Also called angio-osteohypertrophy syndrome
- Characterised by nevus flammeus (portwine stain), venous malformations, lymphatic malformations and soft tissue hypertrophy of the affected limb.
- Can also have large arteriovenous malformations.
- Patients can complain of pain heaviness in the limbs, difficulty in walking due to abnormal length of the limb.
- Excision of veins, sclerotherapy, laser treatment for port-wine stain are few methods to treat the veins.

KEY BOX 10.3

SUPERFICIAL SYSTEM— SALIENT FEATURES

- As the name suggests, they are in the **superficial fascia** and are often visible (saphenous means easily seen).
- **They are low pressure and poorly supported system.**
- They are provided with **numerous valves.**
- The middle coat of these veins consists mostly of **smooth muscle.**
- The **middle coat** is also thicker than that of other veins.
- Normal blood flow is from **superficial to deep system** of veins.

SECONDARY VARICOSE VEINS

- Women are more prone for varicose veins because of the following reasons:
  - **Pregnancy and pelvic tumours** cause proximal obstruction to the blood flow.
  - **Pills** (oral contraceptive pills) alter the viscosity of blood.
  - **Progesterones dilate vessel wall.**
- **Congenital arteriovenous (AV) fistula** increases blood flow and increases venous pressure.
- Deep vein thrombosis can occur as a result of road traffic accidents, postoperatively, etc. This can lead to **destruction of valves** resulting in varicose veins.

SURGICAL ANATOMY OF THE VENOUS SYSTEM OF LEG

It can be discussed under the following headings:

1. Superficial system—long and short saphenous veins and their tributaries
2. Perforators
3. Deep system of veins

SUPERFICIAL VENOUS SYSTEM

Anatomy of the long saphenous vein (LSV)

It starts in the foot from the tributaries of dorsal venous arch, permits reverse flow through its competent valves, ascends in front of medial malleolus and runs along the medial side of the leg. It then ascends in the thigh and ends at the saphenofemoral junction (SF) by joining the femoral vein, which is 1½ inches (4 cm) below and lateral to the pubic tubercle. It has 15 to 20 valves. Absence of valves results in varicose veins (Key Box 10.3 and Fig. 10.1).

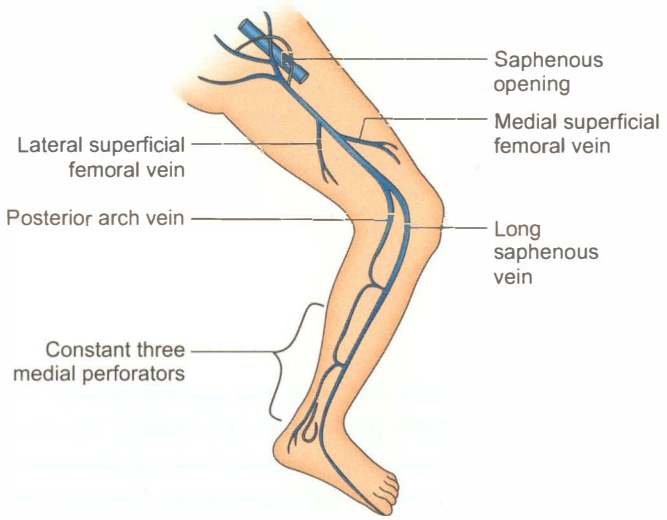


Fig. 10.1: Long saphenous vein, tributaries, communicating veins

Tributaries

Tributaries near the termination

1. Superficial circumflex iliac vein
2. Superficial epigastric vein
3. Superficial external pudendal vein

Tributaries in the lower thigh

1. Lateral superficial femoral vein
2. Medial superficial femoral vein
3. Transverse suprapatellar vein
4. Transverse infrapatellar vein

Tributaries in the leg

1. Anterior vein of the leg
2. Posterior arch vein lies parallel to and behind the main trunk of long saphenous vein. It anastomoses with small venous arches connecting the medial perforating veins.

These tributaries connect the long saphenous with short saphenous veins (SSV). They are also called communicators.

Perforators (Fig. 10.2)

These are the veins which connect long saphenous vein with deep system of veins. Since they perforate deep fascia, they are called perforators. There are 5 constant perforators in the lower limb on the medial side.

- **Leg perforators:** They are 3 in number. The lowest perforator is situated below and behind the medial malleolus. The middle perforator is 10 cm above the tip of the medial malleolus. The upper perforator is 15 cm above the medial malleolus.
- **Knee perforator:** It is situated just below the knee.
- **Thigh perforator:** It is situated a palm-breadth above the knee (Fig. 10.2).

A knowledge of perforators forms the basis of **multiple tourniquet test**. Most of the perforators are provided with valves. Weakness of these valves or damage to valves results in varicosity.

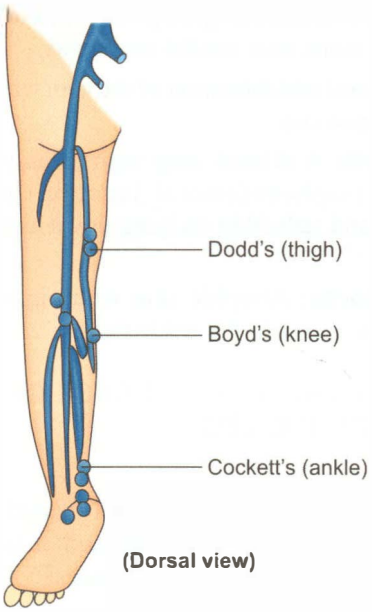


Fig. 10.2: Perforators

Deep venous system

This comprises the femoral and the popliteal veins, veins or venae comitantes accompanying anterior tibial, posterior tibial and peroneal arteries and valveless veins draining the calf muscles (soleal venous sinus).

Salient features of deep venous system

- It is a high pressure system, well-supported by powerful muscles.
- They are connected to superficial veins by means of perforators.
- It is the powerful calf muscle contraction that returns the blood to the heart.
- The deep veins are also provided with valves (Fig. 10.3).

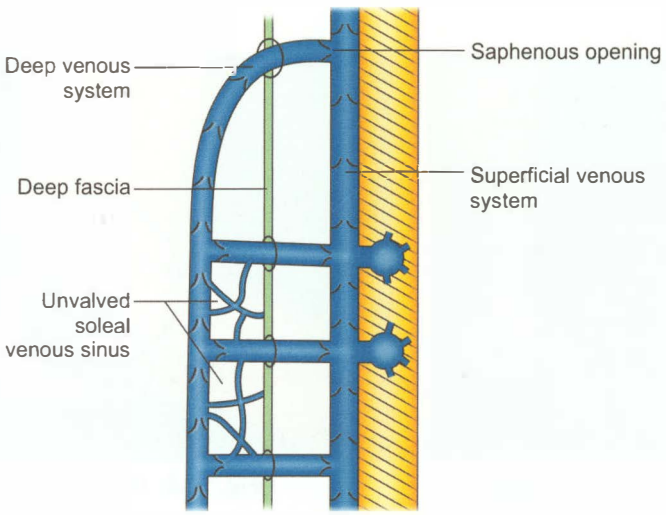


Fig. 10.3: Deep venous system

SURGICAL PHYSIOLOGY

Blood is returned to the heart from the lower limbs by the following mechanisms:

- **Calf muscle pump:** It is the alternate contraction and relaxation of the muscles of the leg (the major factor). The pressure within the calf compartment rises to 200–300 mmHg during walking.
- **Competent valves** (unidirectional) in the leg (Key Box 10.3): When these valves are absent or weak, perforator incompetence develops resulting in varicose veins.
- ***Vis-a-tergo*** of the circulation, that is the pressure transmitted from the arterial tree passes the capillary bed to the venous side. This helps in return of blood to heart at resting position.
- Negative intrathoracic pressure
- Venae comitantes.

KEY BOX 10.4

DISTRIBUTION OF VALVES

- Inferior vena cava : No valve
- Common iliac vein : No valve
- Long saphenous vein : 10–14 valves
- Short saphenous vein : 1 valve

A FEW TERMINOLOGIES FOR DILATED VEINS

1. **Telangiectasia:** It means a confluence of dilated intradermal venules of < 1 mm in diameter.
2. **Reticular veins:** Dilated subdermal veins of 1–3 mm in diameter.

3. **Ankle flare:** Dilated group of reticular (corona phlebectasia) veins near medial malleolus.
4. **Blow out:** A localised dilatation of the vein which is 'dome' like/or ballooned out.
5. **Saphena varix:** A dilated long saphenous vein near the termination (saphenofemoral junction) in the groin which is soft and reducible on lying down and elevation of leg.
6. **Atrophic blanche:** Atrophic skin with pigmentation and reticular veins—seen in the ankle region.

CLINICAL EXAMINATION OF A CASE OF VARICOSITY OF THE LEG

Symptoms

- Majority of the patients present with dilated veins in the leg. They are minimal to start with and at the end of the day they are sufficiently large because of the venous engorgement.
- **Dragging pain** in the leg or dull ache is due to heaviness. Night cramps occur due to change in the diameter of veins. Aching pain is relieved at night on taking rest or elevation of limbs.
- Sudden pain in the calf region with fever and oedema o the ankle region suggests deep vein thrombosis (DVT) Some patients with DVT may be asymptomatic.
- Patients can present with ulceration, eczema, dermatitis an bleeding.
- Symptoms of pruritus/itching and skin thickening.

PEARLS OF WISDOM

Interestingly, pain due to varicose veins is relieved on exercise in contrast to pain due to arterial diseases, which get worse on exercise.

Signs

Inspection (should be done in standing position)

- **Dilated veins** are present in the medial aspect of leg anc the knee. Sometimes they are visible in the thigh also (*see* next page for clinical classification) (Figs 10.4 to 10.10).
- Single dilated varix at SF junction is called saphena varix. It is due to **saccular dilatation** of the upper end of long saphenous vein at the saphenous opening (Tables 10.1 and 10.2).

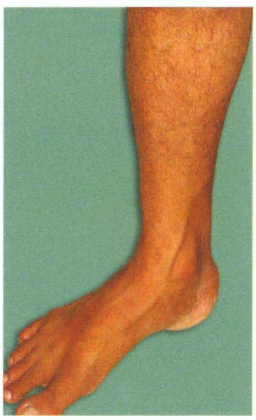


Fig. 10.4: Grade 0

Fig. 10.5: Grade 1

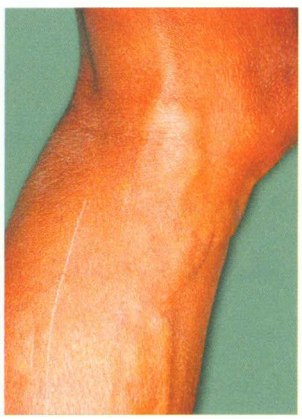


Fig. 10.6: Grade 2

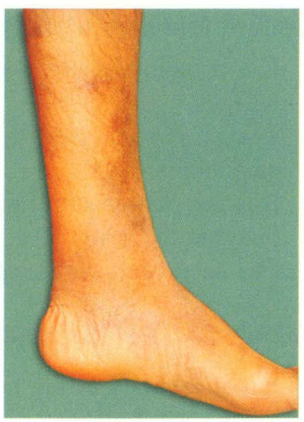


Fig. 10.7: Grade 3

Fig. 10.8: Grade 4

Fig. 10.9: Grade 5

Fig. 10.10: Grade 6

See Table 10.2 for description of various grades of chronic lower extremity venous disease

Table 10.1 Classification[1] of chronic lower extremity venous disease

Classification	Definition
C	Clinical signs (grade 0–6): A for asymptomatic or S for symptomatic
E	Etiologic classification (congenital, primary or secondary)
A	Anatomic distribution (superficial, deep or perforator, alone or in combination)
P	Pathophysiologic dysfunction (reflux or obstruction, alone or in combination)

[1]This classification is from International Consensus Committee on Chronic Venous Diseases.

Table 10.2 Clinical classification of chronic lower extremity venous disease (Figs 10.4 to 10.10)

Grade	Characteristics
0	No visible or palpable signs of venous disease
1	Telangiectases, reticular veins, or malleolar flare
2	Varicose veins
3	Oedema without skin changes
4	Skin changes ascribed to venous disease (e.g. pigmentation, venous eczema, or lipodermatosclerosis)
5	Skin changes as defined above with healed ulceration
6	Skin changes as defined above with active ulceration

- Veins are **tortuous and dilated**.
- A localised, dilated segment of the vein, if present, is an indication of a **blow out**. It signifies underlying perforator (Fig. 10.11).
- **Ankle flare** is a group of veins near the medial malleolus.
- Complications such as ulceration, bleeding, eczema and dermatitis may be present.
- Healed scar indicates previous ulceration.
- Look at the popliteal fossa region also (Fig. 10.12).

PEARLS OF WISDOM

Smaller varicosities are called reticular veins. They are usually 1–3 mm in diameter.

Palpation

- First palpate along the whole length of vein. Look for tenderness. If present, it indicates thrombophlebitis.
- Vein which is thrombosed will feel as a firm/hard nodule.
- Following tests are done (Key Box 10.5).

KEY BOX 10.5

TESTS FOR VARICOSE VEINS AND INFERENCE

- Cough impulse test: SF incompetence
- Trendelenburg I: SF incompetence
  Trendelenburg II: Perforator incompetence
- Multiple tourniquet test: Site of perforator incompetence
- Schwartz test: Superficial column of blood
- Modified Perthes' test: Deep vein thrombosis
- Fegan's test: To locate the perforators in the deep fascia.

1. **Cough impulse test (Morrissey's test):** This test should be done in the standing position. The examiner keeps the finger at SF junction and asks the patient to cough. Fluid thrill, an impulse felt by the fingers, is indicative of *'saphenofemoral incompetence'*.
2. **Trendelenburg test:** This test is done in 2 parts (Figs 10.13 to 10.15).
   - ***Method:*** The patient is asked to lie on the couch in the supine position. The leg is elevated above the level of heart and the vein emptied. SF junction is occluded with the help of the thumb (or a tourniquet) and the patient is asked to stand.

Fig. 10.11: Classical blow out

Fig. 10.12: Dilated vein in popliteal region

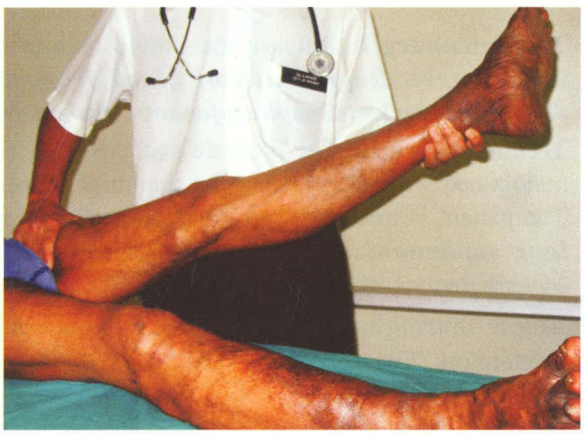


Fig.10.13: Trendelenburg test Part I

Fig. 10.14: Rapid filling on releasing pressure

Fig. 10.15: Trendelenburg test Part II—slow filling is seen; **Figs 10.16A and B:** Multiple tourniquet test (3 or 4)

Trendelenburg I: Release the thumb or tourniquet immediately. ***Rapid gush of blood from above downwards indicates saphenofemoral incompetence.***

- **Trendelenburg II:** The pressure at the SF junction is maintained without releasing the thumb or tourniquet. The patient is then asked to stand. ***Slow filling of the long saphenous is seen. It is due to perforator incompetence*** (retrograde flow of blood).

3. **Multiple Tourniquet test:** It is done to find out exact site of perforators (Fig. 10.16).
   - ***Method:*** The patient is asked to lie supine on the couch The vein is emptied by elevation. As the name suggest 3–5 tourniquets (multiple) can be applied. However, more tourniquets are applied, the exact localisation c the perforators can be made out but it is not practica **There are mainly ankle, knee and thigh perforator** Hence, four tourniquets can be applied at various level as mentioned below.
   - **1st Tourniquet:** At the level of saphenofemoral junctio (SF junction).
   - **2nd Tourniquet:** At the level of middle of the thigh, t occlude perforator in the Hunter's canal.
   - **3rd Tourniquet:** Just below the knee.
   - **4th Tourniquet:** Palm breadth (lower third of the leg above medial malleolus/ankle.

   Ask the patient to stand and observe appearance of veins

   Inference: Appearance of veins between first and secon tourniquets indicates incompetence of thigh perforators between second and third indicates incompetence of kne perforators and below the fourth tourniquet indicate incompetence of ankle perforators. Most commonly, below knee and ankle perforators are incompetent.

   On releasing the tourniquets one by one from below upwards, sudden retrograde filling of the veins occurs.

4. **Schwartz test:** It is done with the patient in the standing position. Place the fingers of the left hand over a dilated segment of the vein and with the right index finger tap the vein below. A palpable impulse suggests a superficial column of blood in the vein and it also suggests incompetence of the valves in between the segment of the vein (Fig. 10.17).

Fig. 10.17: Schwartz test (should be done in the standing position)

5. **Modified Perthes' test**[1]**:** It is done to rule out deep vein thrombosis. The patient is asked to stand, the tourniquet is applied at SF junction and he is asked to have a brisk walk.

   Inference: If the patient complains of severe pain in calf region or if superficial veins become more prominent, it is an indication of **deep vein thrombosis** and is a contraindication for surgery.

PEARLS OF WISDOM

Please note: Vein is not emptied in Perthes' test.

[1]In original Perthes' test, the limb is wrapped and elastic bandage is applied.

6. **Fegan's method (test):** It is done to detect the site of perforators. The patient is asked to stand. The varicosity is marked with methylene blue and he is asked to lie down. The leg is elevated to empty the vein and the vein is palpated throughout its course. The defects in the deep fascia have a circular, buttonhole consistency.

Examination of varicose ulcer: It should be done by inspection and palpation.

Evidence of deep vein thrombosis: Homan's test and Moses' test *(vide infra)* must be done in chronic DVT.

Examination of the abdomen: To rule out pelvic tumours. Look for inferior vena caval obstruction in the form of dilated veins in the lateral abdominal wall.

CLINICAL DISCUSSION

At the end of clinical examination, you should be ready to answer following questions:

1. **Which system is involved?**
   - Medial veins—LSV—long saphenous vein
   - Lateral veins—SSV—short saphenous vein
2. **Is SF junction incompetent?**
   Yes—Trendelenburg I is positive
   No—Trendelenburg I is negative
3. **Is there perforator incompetence?**
   Yes—Trendelenburg II is positive
   No—Trendelenburg II is negative
4. **Which group of perforators are incompetent?**
   According to the results of multiple tourniquet test, mostly ankle and below knee
5. **Is there deep vein thrombosis?**
   Yes—Perthes' test is positive
   No—Perthes' test is negative
6. **Is there any abdominal mass?**
   Pelvic tumours
7. **Any complications are present?**
   Eczema, dermatitis, ulcer, etc.
8. **Is it unilateral or bilateral?**
   Use hand-held Doppler

Investigations

1. **Hand held Doppler** is the first, minimum level investigation to be done before treating a patient with venous disease.
   - When the blood flows, the **wave** emits a signal — the Doppler signal.
   - If there is SF incompetence, forward and backward flow can be detected.
   - **Biphasic signal:** Gently squeeze calf muscles and the flow can be assessed by Doppler probe in the SF junction.
   - It can also pick up accessory long saphenous vein in the groin.
   - It is not an investigation to identify the perforators (Key Box 10.6).

KEY BOX 10.6

DOPPLER ULTRASOUND

- This investigation is carried out with the patient standing/sitting.
- Incompetence of SFJ and saphenopopliteal junction (SPJ) can be assessed by this method—reflux.
- **Gentle squeezing** of calf muscles helps in detecting saphenopopliteal incompetence.
- To identify and locate perforators.
- It also helps to **rule out arterial** diseases.
- It can detect **patency of veins**.
- It can detect deep vein thrombosis.

2. **Duplex ultrasound imaging:** In this investigation high resolution B-mode ultrasound imaging and Doppler ultrasound are used (Figs 10.18 to 10.20).

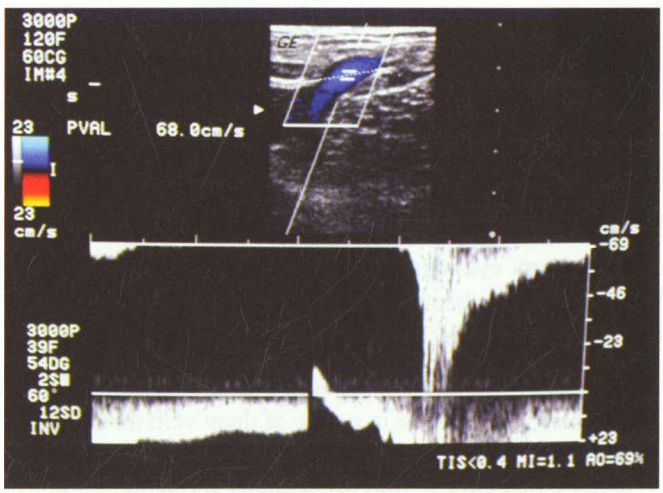


Fig. 10.18: Duplex scanning showing venous reflux

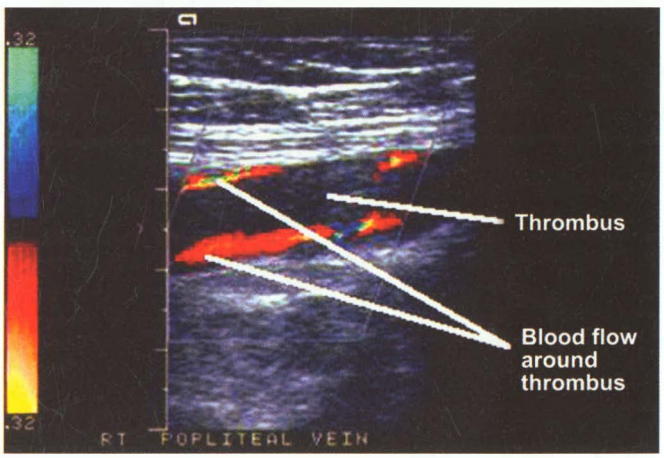


Fig. 10.19: Popliteal vein thrombosis
(*Courtesy:* Dr Rajgopal, Head, Department of Radiology and Imaging, KMC,Manipal)

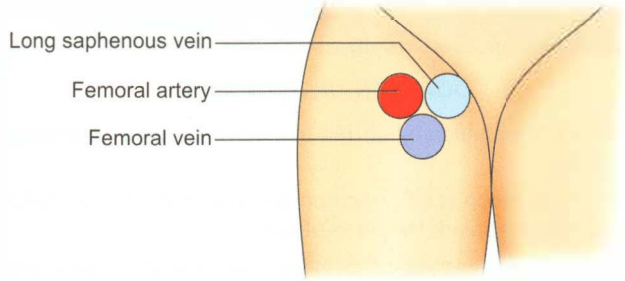


Fig. 10.20: Mickey mouse sign—as seen in duplex ultrasound

- It helps in getting images of veins, measure flow in all lower limb veins.
- Origin of venous ulcers and varicose veins can also be assessed.

PEARLS OF WISDOM

Retrograde flow in the veins can be demonstrated by compression, release and valsalva manoeuvre.

Few 'signs' have been described in the duplex scanning (Fig. 10.20).

'Mickey mouse' sign: The three prominent structures—common femoral artery and great saphenous vein above and common femoral vein below **mimic Mickey mouse** in duplex imaging.

Saphenous eye sign: In the fascial compartments of the thigh, dilated saphenous vein in cross section resemble an 'eye'—in transverse 'B' mode scan.

3. **Venography:** Both ascending and descending venographies can be done in a case of deep vein thrombosis. It is an invasive procedure and risk of spreading infection and septicaemia is present. Duplex ultrasonography has largely replaced this investigation (rarely done now). Varicography refers to injecting contrast into surface veins (indicated in recurrent varicose veins).

   It can be followed by sclerosant injections.

4. **Plethysmography:** They are based on measurement of volume changes in the leg. By placing a light-emitting diode above medial malleolus and patient performing tip-toe movements, venous recovery time can be measured. This is called photoplethysmography (PPT).

PEARLS OF WISDOM

Venous recovery time is shortened in chronic venous insufficiency.

With the advent of venous duplex scan, venography and plethysmography are rarely done.

Treatment

5 types of treatment are popular:

1. Elastic compression stockings
2. Injection line of treatment
3. Foam sclerotherapy
4. Endovenous laser ablation
5. **R**adio-**F**requency **A**blation (**RFA**)

1. Elastic compression stockings

Elastic compression bandage and elevation of leg. It forms the fundamental steps in treating varicose veins. This can be advised in asymptomatic cases of varicose veins and in secondary varicose veins.

- Usually 20–30 mmHg stockings are sufficient.
- It should be worn from ankle to below knee.
- It should be worn during working hours (entire day).
- Should be removed while lying down but legs should be kept elevated.
- ***Above knee stockings should never be prescribed as they are difficult to put on and tend to roll down.***
- *Unna boot:* It is a three-layered dressing.

  Inner layer: It contains roller gauze bandage impregnated with calamine, zinc, glycerine, etc. with graded compression.

  Middle layer: 4″ wide continuous gauze dressing

  Outer layer: Elastic wrap with graded compression.
- Pneumatic compression devices not commonly used.

Indications

- Pregnancy, pelvic tumour
- Perthes' test—positive patient
- AV fistula

2. Injection line of treatment (compression sclerotherapy) (Key Box 10.7)

Indications

1. Below knee varicosity.
2. Recurrent varicosity after surgery.
   - Varicose veins are **marked** in the standing position. The veins are punctured with a needle attached to a syringe containing sclerosant agent and the patient is asked to

KEY BOX 10.7

COMPRESSION SCLEROTHERAPY

- **E**ndothelial cells in the vein wall are damaged
- **E**ffective sclerosant
- **E**mpty vein
- **E**lastic compression stockings
- **E**xercises *Observe* **5 Es**

lie down. **3% sodium tetradecyl sulphate or 1–2 ml of ethanolamine oleate or hypertonic saline** is injected into the column of vein. **Aseptic thrombosis** occurs and when fibrosis occurs, the vein shrinks. Tight elastic compression bandage is applied. Success of sclerotherapy depends upon **effective sclerosant**, injection into an **empty vein** and **compression followed by exercise**.

PEARLS OF WISDOM

Injection sclerotherapy is useful in varicose veins less than 3 mm in diameter. Large veins require surgery.

Complications of sclerotherapy

- Allergy
- Pigmentation
- Deep vein thrombosis
- Thrombophlebitis
- Skin necrosis

3. Ultrasound-guided foam sclerotherapy (Key Box 10.8)

- It is popular in a few centres in the UK only.
- Foam sclerosant C (Polidocanol) is used. It is prepared by air mixing technique with sclerosant.
- It is injected into veins (superficial) under ultrasound guidance.
- LSV should be compressed in the upper thigh to prevent foam from entering into femoral vein.
- Elevate the leg to prevent foam from entering into calf veins.

Complications of foam sclerotherapy

- Extravasation: Skin ulceration
- Escape into deep veins: Deep vein thrombosis
- Entering into brain (air): Headache and stroke.

KEY BOX 10.8

FOAM SCLEROTHERAPY

- Used to treat small and medium sized varicose veins.
- Sclerosant is mixed with air or preferably carbon dioxide plus oxygen.
- Carbon dioxide plus oxygen is soluble in blood and hence safe.
- Foam causes inflammation of vein wall, obliteration of venous lumen and vein occlusion.
- Success rates range from 60 to 90%.
- Complications include cutaneous ulceration, thrombophlebitis, deep vein thrombosis and allergy.
- Foam displaces blood (unlike sclerosant which mixes with blood), requires small quantity to be in touch with vein wall.
- Extravasated foam is much better tolerated.
- It is echogenic.

4. Endovenous laser ablation (EVLA)

Introduction: It is a minimally invasive, outpatient procedure using laser fibre to ablate varicose veins. This gives excellent cosmetic and functional results.

Procedure (Fig. 10.21)

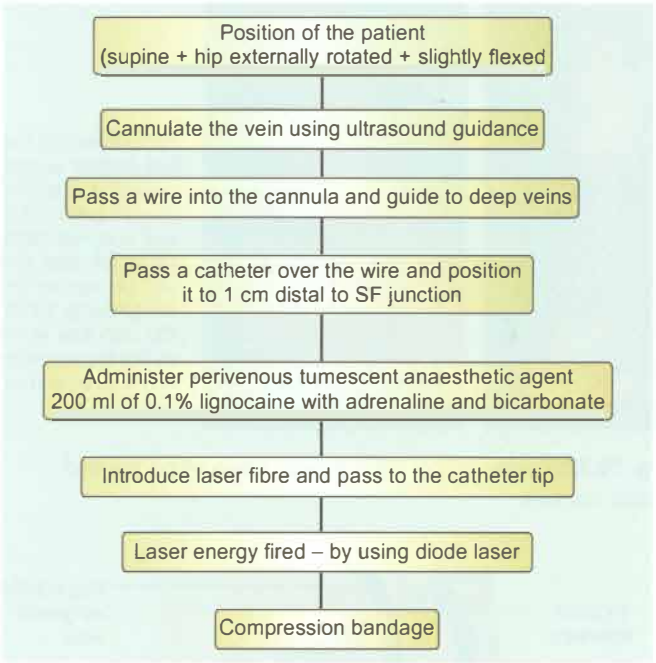


Fig. 10.21: Endovenous laser ablation

Advantages

- Simple, outpatient procedure
- Ideal for junctional and truncal incompetence
- Less expensive than radiofrequency ablation.

Disadvantages

- Not ideal for smaller veins, tortuous veins and thrombophlebitic veins
- Costly.

5. Radiofrequency ablation (RFA)

- It is also a minimally invasive technique of ablation of the veins by using a bipolar catheter at a temperature of 85–120°C, using a power of 2–4 W.
- The position of the patient, accurate marking of the veins, tumescent anaesthesia, are similar to that of EVLA.
- However, the vein is cannulated with a 7F sheath and catheter is not positioned within 2 cm of the SF junction.
- Segment of the vein over 7 cm long can be ablated.
- Complications include thrombophlebitis, pain, skin burns.

6. Surgery

1. Trendelenburg's operation (Figs 10.22 to 10.27)

- An inguinal incision is made, long saphenous vein identified and the 3 tributaries are ligated. Long saphenous vein is ligated close to the femoral vein ***juxtafemoral flush ligation***.

Fig. 10.22: Perforators marked

Perforators are marked before surgery to facilitate identification of perforators and communicating veins so that they can be ligated and excised (Fig. 10.22). You can see in this picture the prominent veins are marked (Fig. 10.23).

Fig.10.23: Ankle veins marked

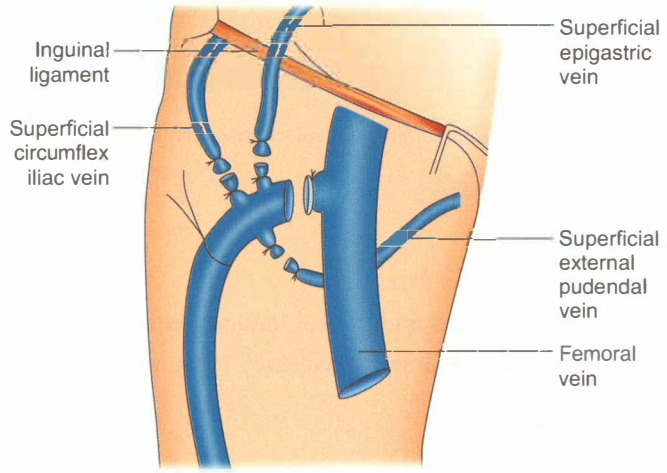


Fig. 10.24: Trendelenburg operation. Long saphenous vein should never be ligated in the groin **unless you demonstrate 'T' junction**—LSV joining femoral vein (Key Box 10.9)

KEY BOX 10.9

WISDOM IN JUXTAFEMORAL FLUSH LIGATION

1. Demonstrate T, junction—Long saphenous vein joining femoral vein.
2. Demonstrate 3–4 tributaries before ligation.
3. Double saphenous vein can be present as an anomaly.
4. Receives large anterolateral thigh veins and posteromedial thigh veins.
5. Demonstrate femoral vein for a centimetre both above and below the SF junction.

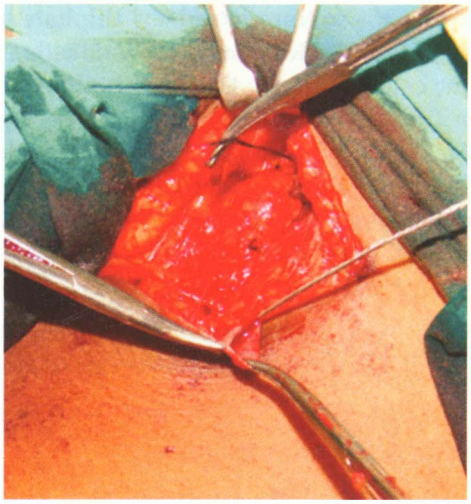


Fig. 10.25: LSV is divided at the groin and stripper is introducec within the lumen

Fig. 10.26: One segment of the vein is isolated followed by ligation at both ends and if it is very tortuous, it is excised

Fig. 10.27: Multiple ligation

- An incision is given on the medial side of the upper leg and long saphenous vein is isolated. The lower end is ligated and the vein incised. A long metallic stripper is

introduced within the vein and brought out from the long saphenous vein below knee incision. A metallic head is connected to the stripper and the vein is avulsed. Tight crepe bandage is applied, inguinal incision sutured and the limb is elevated (retrograde avulsion).

- During the procedure, **the perforators get avulsed** from the long saphenous vein and get thrombosed. Hence, this procedure is called ligation with stripping operation. It is indicated in cases of saphenofemoral incompetency.
- Once stripping is done, small incisions are given where the tributaries are marked. The vein is cleared of subcutaneous tissue and is avulsed.
- Carefully isolate a long segment of remaining vein and avulse it.

2. Subfascial ligation of Cockett and Dodd

- In this operation, the perforators are identified deep to deep fascia and are ligated subfascially. This is indicated in cases of perforator incompetence with saphenofemoral competence. This can also be done using an endoscope.

3. Subfascial endoscopic perforator surgery (SEPS) (Fig.10.28)

- **Small port incisions** are made in the skin of calf region, deepened through the fascia.
- **Carbon dioxide** insufflation is done. A balloon expander may also be used to distend the subfascial plane.

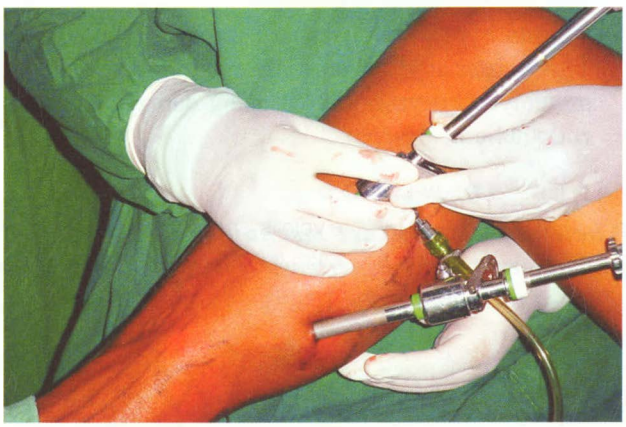


Fig. 10.28: SEPS

- **2–6 perforators** are identified and ligated.
- The procedure is simple, quick with least morbidity and is becoming popular.
- Indicated for below knee perforators.

Recent techniques for the management of varicose veins

1. VNUS closure

- By using ultrasound control, an ablation catheter is inserted into saphenofemoral junction and slowly withdrawn.
- In this process, veins are destroyed.
- This procedure has the advantage of lesser incidence of thigh haematoma and pain.
- Complications include deep vein thrombosis, recurrence and damage to overlying skin.

2. TriVex

- In this method, veins are identified by subcutaneous illumination, followed by injection of large quantities of fluid.
- Then superficial veins are sucked out.
- Complications include induration, bruising and subcutaneous grooves.

Complications of varicose veins

1. **Eczema and dermatitis:** It occurs due to extravasation and breakdown of RBCs in the lower leg. It gives rise to itching which precipitates varicose ulcer. It is treated by application of zinc oxide cream or silver sulfadiazine cream (stasis dermatitis) (Figs 10.29 and 10.30).
2. **Lipodermatosclerosis** (Fig. 10.31) refers to various skin changes in the lower leg associated with varicose veins such as **thickening of subcutaneous tissue, indurated wood like feel,** pigmentation, etc. It is due to increased venous pressure resulting in capillary leakage with extravasation of blood and fibrin into surrounding tissues. Blood is broken down and haem is released. This combines with iron giving rise to haemosiderin which is responsible for pigmentation. Classically this affects gaiter area of leg just above the malleoli (Figs 10.30 and 10.31).

Fig. 10.29: Eczema

Fig. 10.30: Dermatitis

Fig. 10.31: Observe pigmentation, lipodermatosclerosis and ulcer formation

3. **Haemorrhage:** It occurs due to trauma or eczema. It can be controlled by elevation of the leg and crepe bandage application. Do not try to catch bleeders with artery forceps.

4. **Thrombophlebitis:** It refers to inflammation of a superficial vein. The vein is tender, hard and cord-like. The skin is inflamed and pyrexia is usually present. It is treated by bedrest, elevation, crepe bandage, antibiotics and anti-inflammatory drugs (Key Box 10.10).

KEY BOX 10.10

THROMBOPHLEBITIS—CAUSES

- Spontaneous TAO, malignancy
- Trauma
- Blood transfusion
- IV fluids, chemotherapeutic drugs
- Varicose veins

5. **Venous ulcer:** It is also called **gravitational ulcer** (Figs 10.32 to 10.35). Precipitating factors are **venous stasis** and **tissue anoxia**. *Deep vein thrombosis* is also an important cause of venous ulcer wherein valves are either destroyed or incompetent due to damage. **Sustained venous pressure** results in extravasation of cells, activation of capillary endothelium resulting in **release of free radicals**. These free radicals cause tissue destruction and ulceration. **Lipodermatosclerosis, tissue anoxia** are the other factors. Following hypotheses may explain genesis of varicose ulcers.

Fibrin cuff hypothesis

- The combination of capillary proliferation and inflammation in the form of presence of macrophages is a major factor in the development of venous ulcer. As a result of chronic inflammation, perivascular cuff develops around the capillaries. This perivascular cuff is made up of many connective tissue proteins including fibrin, collagen IV and fibronectin. Slowly, venous ulcer develops.

Fig. 10.32: A case of lipodermatosclerosis

Fig. 10.33: Varicose vein with venous ulcer (*Courtesy:* Dr Maruthu Pandyan, Government Medical College, Madurai, Tamil Nadu)

White cell trapping hypothesis

- Venous hypertension causes trapping of leucocytes. These leucocytes become activated and release proteolytic enzymes thus causing damage to capillary endothelium.

PEARLS OF WISDOM

Whatever be the exact mechanism of ulceration, ambulatory venous hypertension is the only accepted cause of ulceration (Fig. 10.34).

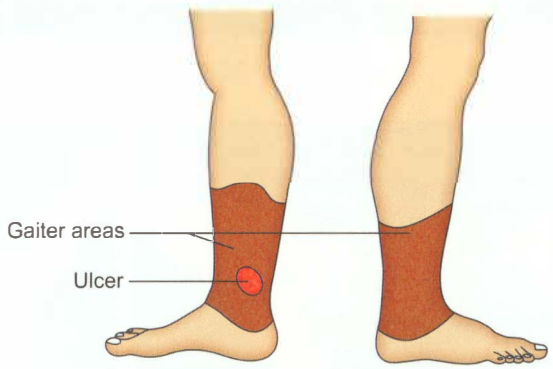


Fig. 10.34: **Gaiter** areas are those areas where **skin changes** and **venous stasis ulcer occurs** due to **ambulatory venous hypertension** and incompetent perforators. Damage to capillaries results in protein-rich fluid and blood extravasation

Fig. 10.35: Venous ulcer (*Courtesy:* Dr Maruthu Pandyan, Prof of Surgery, Government Medical College, Madurai, Tamil Nadu)

Features of venous ulcer (Fig. 10.35)

- Typically, ulcers are situated just above the medial malleolus.
- The ulcers are oval, small, painless, superficial with pigmentation all around.
- Dilated veins above the ulcer give the clue to the diagnosis (Key Box 10.11).

KEY BOX 10.11

FACTORS PREDISPOSING TO NONHEALING VENOUS ULCERS

1. Ambulatory venous hypertension.
2. Perivascular fibrin cuff resulting in poor diffusion of oxygen to the tissues.
3. White cell trapping.
4. Reactive oxygen species are increased and they generate free radicals leading to tissue damage.
5. Inhibition of growth factors leading to poor repair.

Treatment of venous ulcer: *Bisgaard's method*

- Rest with **elevated limb** (Fig. 10.36).
- **Elastic crepe bandage** helps in venous return.
- **Active exercises** should be taught to the patients (to contract calf muscles).
- **Passive exercises**
- Correct way of walking with the **heel down first**.
- If the ulcer is infected, antibiotics are given and dressing of the ulcer is done. Once the ulcer heals, Trendelenburg's operation is done.

6. **Calcification** can be seen in the walls of the vein.
7. **Periostitis** of the tibia can occur due to the ulcer situated on the medial surface of the leg. Due to involvement of the periosteum, the ulcer gives rise to severe pain.
8. **Equinovarus deformity** occurs due to improper habit of walking on the toes which results in shortening of the tendo-Achilles.

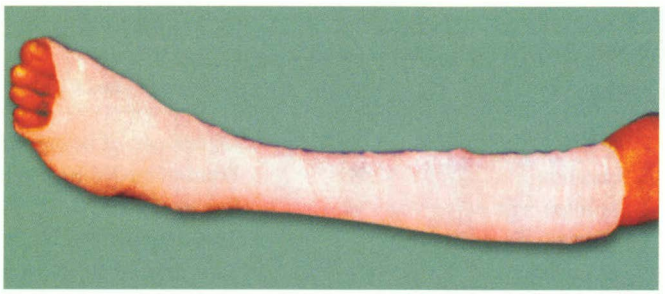


Fig. 10.36: Elastic crepe bandage for varicose ulcer. Elevation of the leg is the most important factor for healing of the ulcer

TEN COMMANDMENTS OF TREATMENT OF VENOUS ULCERS

1. Should take rest with leg elevated above heart level
2. Should receive antibiotics if infection is present
3. Should apply wound cover with suitable dressings
4. Should apply compression stockings
5. Should do venous Doppler to rule out deep vein thrombosis or thrombophlebitis
6. Should consider surgery only when infection is under control
7. Should ligate long or short veins if they are incompetent
8. Should ensure that all perforators are ligated
9. Should look for accessory long saphenous vein near the termination of the vein—if present it should be ligated, otherwise definite chance of recurrence exists
10. Should loose weight

9. **Marjolin's ulcer** is a squamous cell carcinoma arising from healed varicose ulcer with scarring.

SHORT SAPHENOUS VARICOSITY

They are uncommon causes of varicosity in the leg. Short saphenous vein originates from the lateral part of the dorsal venous arch and ends in the popliteal vein in the popliteal fossa (Fig. 10.37). Incompetence of saphenopopliteal valve results in short saphenous varicosity. It produces prominent veins on the lateral aspect of the leg with or without ulceration. They are treated by ligation of the short saphenous vein in the popliteal fossa (Table 10.3).

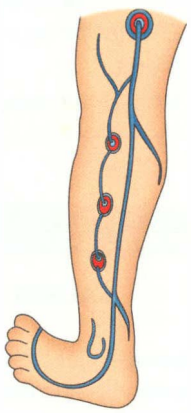


Fig. 10.37: Short saphenous vein and its perforators

Ligation of saphenopopliteal junction

- Preoperative ultrasonographic marking is very essential.
- Vein should be ligated deep to deep fascia.
- Branches—**Giacomini vein** and **gastrocnemius** veins may be seen. They must be ligated.

Table 10.3 Comparison between long saphenous and short saphenous veins

Features	Long saphenous vein	Short saphenous vein
1. Origin	Medial part of dorsal venous arch	Lateral part of dorsal venous arch
2. Location	Front of medial malleolus	Behind lateral malleolus
3. Relation with nerve	Saphenous nerve	Sural nerve
4. Number of valves	15–20 valves	1 valve
5. Termination	Saphenofemoral junction	Saphenopopliteal junction

- It can be stripped up to mid-calf so as to avoid **injury to sural nerve**.
- It is important to close the deep fascia to avoid unsightly cosmetic bulge behind the knee.

DEEP VEIN THROMBOSIS (DVT)

It is also called phlebothrombosis. It is an acute thrombosis of deep veins. Deep vein thrombosis is very common in the western countries, the exact cause of which is not known. Postoperative immobilisation, pressure on the calf muscles, sluggish blood flow and prolonged bed rest are the various factors which precipitate deep vein thrombosis. Commonly, it affects venous sinuses in the soleal muscles. It is a common starting place. It can also involve pelvic veins. Various factors responsible for deep vein thrombosis can be remembered as THROMBOSIS (Key Boxes 10.12 and 10.13).

KEY BOX 10.12

THROMBOSIS—VIRCHOW'S TRIAD

- Endothelial injury
- Stasis
- Increased coagulability

KEY BOX 10.13

CAUSES OF DEEP VEIN THROMBOSIS (LOWER LIMB DVT)

Trauma—injury to the vessel wall
Hormones—increased coagulability
Road traffic accidents
Operations—cholecystectomy
Malignancy—sluggish blood flow
Blood disorders—polycythaemia
Orthopaedic surgery, obesity, old age
Serious illness—stroke, MI
Immobilisation
Splenectomy

Remember as **THROMBOSIS**

Upper limb DVT is rare. Can occur due to trauma or surgery.

Clinical features

- The maximum incidence occurs on the 2nd day and 5th–6th days in the postoperative period.
- First complaint is usually oedema, erythema, dilated veins of the leg (Figs 10.38 to 10.41).
- Dull-aching or nagging pain in the calf muscles is present.
- Superficial blebs in the skin.
- Low grade fever with increased pulse rate is characteristic.
- Phlegmasia alba dolens refers to white leg. It occurs when the thrombus extends from calf region to iliofemoral vein.
- Phlegmasia coerulea dolens refers to blue leg with loss of superficial tissues of the toes.

Signs (acute DVT)

1. **Homan's test:** Forcible dorsiflexion of foot results in severe pain in the calf region.
2. **Moses' test** (Ideally should not be done for fear of embolism): Tenderness over calf muscle on squeezing the muscle from side to side.

Investigations

1. **Doppler study:** It is ideal for femoral vein thrombosis or when thrombus extends into popliteal vein. Normal femoral vein gives a wind storm sound which completely disappears at the end of inspiration. No sound is heard if there is femoral thrombosis (Key Box 10.14).
2. **Contrast venography:** It is done by injecting radiopaque dye into dorsal venous arch with an inflatable cuff both above the ankle and above the knee. Clot appears as a filling defect. However, venography is not routinely done because it is expensive and invasive.

KEY BOX 10.14

DUPLEX SCANNING IN DEEP VEIN THROMBOSIS (B-MODE)

- Vein is larger than normal because of occlusion.
- Not completely compressible.
- Lacks respiratory variation.
- Does not show flow augmentation with calf compression.
- May have collateral flow.

Fig. 10.38: Swollen right leg—early indication of DVT. It responded to low molecular weight heparin

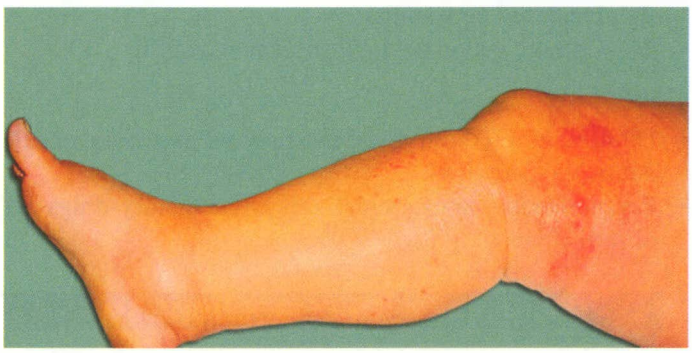


Fig. 10.39: Acute DVT: Grossly swollen, oedematous leg. Skin is red and warm with superficial blisters

Observe the differences between acute DVT and chronic DVT. Such swollen legs have been mistaken for filariasis and treated also without any results.

Fig. 10.40: Swollen leg, pigmentation, eczema and secondary varicosity in a case of chronic DVT

Fig. 10.41: Grossly swollen leg, pigmentation, eczema and secondary infection in a case of DVT

Treatment

1. Bedrest and elevation of limbs
2. **Injection heparin** 10,000 units IV bolus with continuous infusion of 30,000 to 45,000 units per day. During heparin therapy **activated partial thromboplastin time (APTT)** should be maintained at twice the normal value. Heparin is given for a period of 7–10 days.
   - **Warfarin**, an oral anticoagulant is started 2–3 days before heparin is withdrawn because of the slow onset time of warfarin. Treatment with warfarin should continue for 6 to 12 months. Repeat duplex scan should be done to look for recanalisation of the veins. The dose of warfarin should be 10 mg twice a day. **Treatment is monitored with prothrombin time and INR.** International Normalized Ratio (INR) should be between 2.0 and 3.0.
3. **Low molecular weight heparin (LMWH):** It is given once or twice a day, in the form of injection. **No blood monitoring is required**. Incidence of bleeding is less with LMWH.
4. **Inferior vena caval filters:** They can be inserted percutaneously *via* femoral vein in patients wherein **lytic therapy** is contraindicated.

PEARLS OF WISDOM

IVC filters are more commonly indicated in patients with recurrent DVT with symptomatic pulmonary embolism.

5. **Surgery** is not done regularly. However, in chronic cases venous bypass has been attempted with moderate success.
   - **Palma** operation is done in iliofemoral thrombosis wherein common femoral vein below the block is anastomosed to the opposite femoral vein using long saphenous, from the opposite side
   - **May-Husni** operation wherein popliteal vein is connected to long saphenous vein above.

Complications

1. **Permanent oedema** of the limb. The limb has an **inverted beer bottle** appearance.
2. **Pulmonary embolism** because the thrombus is not attached to vessel wall.
3. **Secondary varicosity** and **nonhealing ulcer**.

Prophylaxis of DVT (Key Box 10.15)

KEY BOX 10.15

PROPHYLAXIS: RISK GROUPS

- Low risk : Young patients
  Minor illness
  Operation < 30 minutes
- Moderate risk : > 40 years
  Debilitating illness
  Major surgery
- High risk : > 50 years, medical conditions—MI, stroke, major surgery, malignancy, obesity.

- Decrease obesity and advise exercises before surgery.
- **Low dose heparin** 5,000 units subcutaneous, 2 hours before surgery and 24 hours after surgery, and then every 12 hours for 5 days is given, during major surgeries such as cholecystectomy, abdominoperineal resection, etc.
- **Intermittent pneumatic compression** of the calf throughout the operation, maintains the blood flow in the lower limbs. Inflation pressure is around 30–50 mmHg.
- Dextran 40 inhibits sludging of red blood cells and platelet aggregation.
- **Aspirin** along with dipyridamole has been used (antiplatelet agents).
- **Early mobilisation, walking, adequate hydration.**
- Low molecular weight heparin decreases chances of bleeding.

MISCELLANEOUS

RECURRENT VARICOSE VEINS

- Incidence is about 10–30%.
- Important causes of recurrence is failure to ligate the LSV at SF junction, failure to ligate tributaries at SF junction, and a possible accessory long saphenous vein (Fig. 10.42).
- Other factors for failure are neovascularisation, reflux in residual veins.

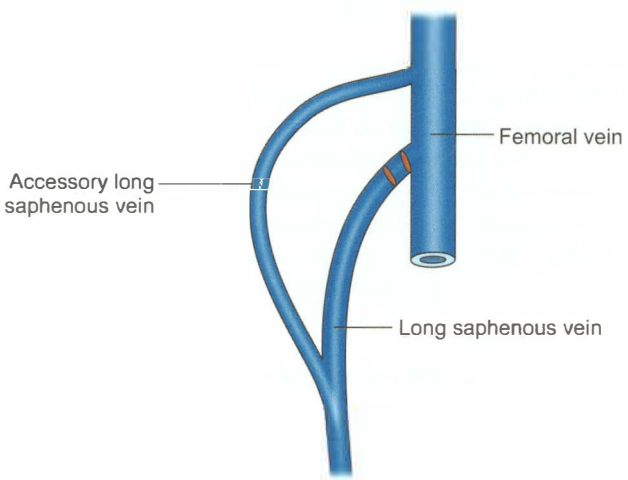


Fig. 10.42: Accessory long saphenous vein

- Recurrence is less after stripping of veins than multipl ligation.
- Injection sclerotherapy and EVLA/RFA can be done.
- Re-surgery can be done. However, wound infection, woun gaping, lymph leak are more common.

PELVIC CONGESTION SYNDROME

Few premenopausal patients complain of dull aching/sever pain in the pelvis. Clinical examination may reveal tenderne in the hypogastrium. A few salient features given below ca be remembered by mnemonic—**PELVIC**

- **P** Premenopausal patients
- **E** Excessive micturition, excessive bleeding during menstruation
- **L** Leg varicosity in atypical sites such as in the thigh
- **V** Varicosity of ovary, vulva, pelvic veins cause for pelvic pain
- **I** Increased on standing
- **C** Chronic pelvic pain, which is noncyclical

Management

- Ultrasound, CT scan and MRI may be necessary to rule out pelvic pathology
- Psychotherapy, nonsteroidal anti-inflammatory drugs are of some help.

PULMONARY THROMBOEMBOLISM

Clinical features of DVT with pulmonary thromboembolism

- Tachypnoea and tachycardia
- Chest pain
- Breathlessness
- ECG may show in S1 Q3 T3 pattern (S wave in lead I and Q and inverted T waves in lead III).

Investigations

- Ventilation perfusion scan (V/Q)
- CT angiography of thorax.

Treatment

- Rest, compression bandage, elevation.
- Anti-DVT treatment

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated.
- New/alternate treatment of varicose veins in the form of endolaser ablation, radiofrequency ablation have been given.
- Ultrasound-guided foam sclerotherapy has been discussed.
- Recurrent varicose vein, pelvic congestion syndrome and new signs of varicose veins in the duplex scan have been added

MULTIPLE CHOICE QUESTIONS

1. Which of the following statements is false in long saphenous vein anatomy?
A. It starts from dorsal venous arch
B. It ascends in front of the medial malleolous
C. It has 15–20 valves
D. Posterior arch vein is not a tributary of long saphenous vein

2. Following are true for short saphenous vein *except*:
A. Has 6 valves
B. It ascends behind lateral malleolous
C. Its termination is not constant often
D. Incompetency results in ulcer on the lateral side of the leg

3. Following are features of deep venous system *except:*
A. High pressure system
B. Has valves
C. Present in the soleal group of muscles
D. Connected to superficial veins

4. Following are true for perforators *except:*
A. They communicate superficial and deep veins
B. They are present in the leg
C. They do not have valves
D. SEPS is the endoscopic surgery done on perforators

5. Trendelenburg test is done to find out:
A. Sapheno-popliteal in competency
B. Sapheno-femoral incompetency
C. Deep vein thrombosis
D. Site of perforators

6. Perthes' test is done to find out:
A. Saphenopopliteal in competency
B. Saphenofemoral incompetency
C. Deep vein thrombosis
D. Site of perforators

7. Characteristic feature of venous ulcer in the leg is:
A. Deep painful ulcer
B. Superficial ulcer with pigmentation around
C. Penetrating ulcer with visible bone
D. Ulcers on the dorsum of the foot

8. Plethysmography is based on measurement of which of the following?
A. Volume changes B. Pressure changes
C. Sound changes D. Velocity

9. Following are true for Doppler ultrasound *except*:
A. It can detect patency of veins
B. It can detect arterial pulses
C. It acts with Doppler principle
D. To visualise movement of blood within vessel

10. Following are true about injection sclerotherapy for varicose vein treatment *except:*
A. Vein has to be full during injection
B. Below knee varicosity can be treated with sclerotherapy
C. Recurrent varicosity can be treated with sclerotherapy
D. Useful in veins with less than 3 mm in diameter

11. About radiofrequency ablation:
A. Long saphenous varicosity can be treated
B. Hepatoma larger than 6 cm are often indication for treatment
C. Can be used for Barrett's oesophagus
D. Nerves and cardiac muscles are not stimulated during the treatment

12. Following are true for venous ulcer formation *except:*
A. Ambulatory venous hypertension
B. Free radical release
C. Fibrin, fibronectin, collagen IV
D. Haem deposition

ANSWERS

1 D 2 C 3 C 4 C 5 B 6 C 7 B 8 A 9 D 10 A
11 B 12 D

11

Skin Tumours

- Classification
- Premalignant lesions
- Basal cell carcinoma
- Squamous cell carcinoma
- Melanocytic tumours
- Malignant melanoma
- Other malignant skin tumours
- Other skin lesions
- What is new? / Recent advances

Introduction

The skin, the outermost coat of the human body, functions as a protective cover against various insulting agents such as ultraviolet radiation of sunlight, excessive heat and various chemical agents.

Hence, no wonder it is one of the commonest cancers in elderly patients. However, more than 90% of skin tumours are curable because they are diagnosed early and easily (unlike intra-abdominal malignancies). Many of them are low grade cancers.

- Among skin cancers, about 70% are basal cell carcinomas, 20% are squamous cell carcinomas and 5% are melanocarcinomas.
- Other rare skin cancers are sebaceous carcinomas, dermatofibrosarcomas, etc. In this chapter, only common malignant skin tumours are discussed.
- Also some common skin lesions such as corn and wart are also discussed in this chapter.

CLASSIFICATION OF SKIN TUMOURS

I. Epidermal tumours

A. Benign

- Papilloma
- Seborrhoeic keratosis
- Verrucous naevus

B. Malignant

- Basal cell carcinoma
- Epithelioma, Marjolin ulcer

II. Melanocytic tumours

A. Benign

- Junctional naevus
- Compound naevus
- Intradermal naevus
- Hutchinson's freckle
- Hairy and blue naevus

B. Malignant

- Superficial spreading melanoma
- Nodular melanoma
- Lentigo maligna melanoma
- Amelanotic melanoma
- Acral lentiginous melanoma
- Desmoplastic melanoma

III. Sweat gland tumours (malignant)

- Hidradenocarcinoma
- Adenoid cystic carcinoma

IV. Sebaceous gland tumours

- Sebaceous adenoma
- Sebaceous carcinoma

V. Other tumours

- Dermatofibrosarcoma protuberans
- Trichofolliculoma (hair follicle tumour)

PREMALIGNANT LESIONS OF THE SKIN

1. **Chronic irritation** to the skin can occur due to various chemicals such as dyes, tar and inorganic arsenic, which contain various carcinogens. Coal tar contains polycyclic aromatic hydrocarbons such as benzopyrenes which are carcinogenic-arsenic, soot, mineral oil.

2. **Solar keratosis (actinic keratosis):** Prolonged exposure to sun rays can cause *hyperkeratosis* of the skin which is called solar keratosis. Skin changes occur due to accumulated effect of ultraviolet rays over a period of many years. Ultraviolet rays are also present in phototherapy given in the treatment of psoriasis (**PUVA** therapy—**P**soralen **U**ltra **V**iolet-**A**). **Common sites:** Back of hands, face, rim of ears.
   - ***Age group:*** Middle age, more than 50 years.
   - Clinically the lesion is an irregular, firm and irritating patch which is flat or raised, better felt than seen.
   - Malignancy should be suspected when the lesion becomes indurated, when a nonhealing ulcer develops, when the central crust is shed or when regional lymph nodes are palpable.

3. **Chronic scar:** Squamous cell carcinoma which develops in a scar tissue is called ***Marjolin ulcer*** (Fig.11.1). **Burns scar** is the commonest cause of Marjolin ulcer followed by scar due to varicose ulcer, snakebite scar, chronic osteomyelitis scar, lupus vulgaris (tuberculosis of face) scar.
   - Marjolin ulcer differs from squamous cell carcinoma by following characteristics (Table 11.1).

PEARLS OF WISDOM

Once Marjolin ulcer infiltrates normal skin, it behaves like squamous cell carcinoma.

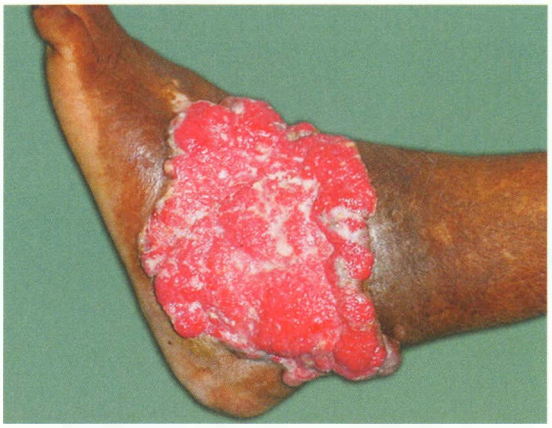


Fig. 11.1: Marjolin ulcer arising in a burns scar

4. **Radiodermatitis:** Increased incidence of skin cancer was found in persons who worked in the radiology department initially. Now, the incidence is less due to the usage of protective gear. Radiation change in the skin may vary from a simple erythema initially to atrophy or hyperpigmentation. Later, this lesion changes into squamous cell carcinoma.

5. **Bowen's disease** is an intraepidermal carcinoma. Chronic solar damage, inorganic compounds and human papilloma virus (HPV 16) are possible aetiological factors. It is rare and occurs in middle-aged patients. It occurs on the skin of the trunk as scaly, erythematous plaques which are often multiple. They are brownish patches with raised margin. Microscopically, large clear cells are found (these cells are also found in Paget's disease of the nipple). Topical application of 5-FU and/or imiquimod is an effective treatment (Figs 11.2 to 11.4).

6. **Leukoplakia**

7. **Autosomal recessive disorders:** In this group, one or more of the DNA repair enzymes are defective or deficient. As a result of this, sites exposed to sun are vulnerable for the development of various skin cancers. **Xeroderma pigmentosum** and **albinism** have increased predisposition to skin cancer (Fig. 11.6).

PEARLS OF WISDOM

White race is a definite risk factor for skin cancer.

Table 11.1 Comparison of Marjolin ulcer and squamous cell carcinoma

Marjolin ulcer	Squamous cell carcinoma
Grows very slowly because of scar tissue	Grows slowly
It is painless as scar does not contain nerves	It can be painful if it infiltrates the nerve fibres
Lymphatic metastasis does not occur because lymphatics are destroyed or occluded	Lymphatic metastasis is the chief method of spread
It is less malignant	Comparatively more malignant
Surgery cures the disease, radiotherapy is not very useful	Both surgery and radiotherapy are used

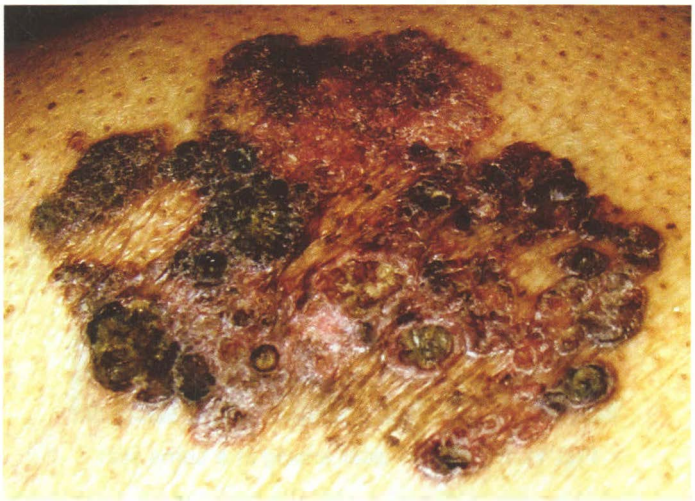


Fig. 11.2: Bowen's disease—premalignant condition

Fig. 11.3: Albinism with squamous cell carcinoma of the right elbow region—patient also has photophobia (*Courtesy*: Dr Satish Deshmukh and Dr Murtaza Akhtar, NKP Salve Medical College and Research Center, Nagpur)

Other Genetic Syndromes

- Muir-Torre syndrome
- Dystrophic epidermolysis bullosa
- Werner syndrome
- Nevoid basal cell carcinoma
- Li-Fraumeni syndrome
- Previous diagnosis of cutaneous carcinoma increases the risk to 35% at 3 years and 50% at 5 years.

BASAL CELL CARCINOMA (RODENT ULCER)

It is the ***most common malignant skin tumour.*** It arises from basal cell of the pilosebaceous adnexa and occurs only on the skin. Generally, it is a slow-growing neoplasm which can present as an ulcer of many years duration. ***Metastasis and death from this disease is extremely rare.*** In some cases, it can present as locally penetrating, ulcerative and destructive lesion.

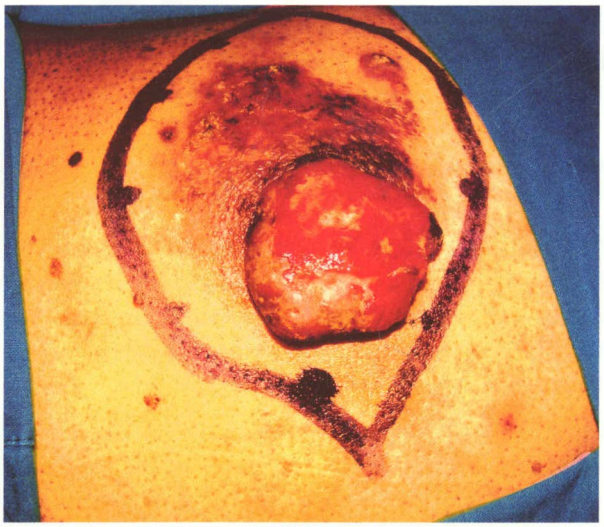


Fig. 11.4: Bowen's disease with ulcerated squamous cell carcinoma

Location

Majority of the lesions are found on the face above a line from lobule of the ear to the angle of mouth.

Common sites (Fig. 11.5)

- Inner canthus of the eye
- Outer canthus of the eye
- Eyelids
- Bridge of the nose
- Around nasolabial fold

These sites are the areas where the tears roll down. Hence it is also called **tear cancer** (Figs 11.6 to 11.8).

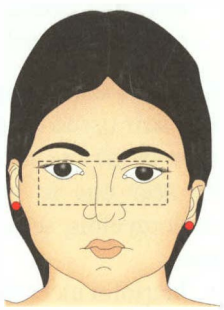


Fig. 11.5: Sites of basal cell carcinoma (BCC)

PEARLS OF WISDOM

Basal cell carcinoma cannot occur in the mucosal surfaces which do not have pilosebaceous adnexa,* e.g. *cervix, lips, tongue.

Precipitating factors

- **Ultraviolet rays:** Basal cell carcinoma is common in Australia[1] and New Zealand because of ultraviolet rays.
- Fair skin is vulnerable for the development of basal cell carcinoma.

[1]This may be the reason why Australian and New Zealand cricket players apply some protective cream to the potentially risky sites while playing the match.

Fig. 11.6: Basal cell carcinoma involving the right ear

Fig. 11.7: Basal cell carcinoma involving nasolabial fold skin

A brother and sister with **xeroderma pigmentosum** developed 33 and 26 skin malignancies respectively which included basal cell carcinoma, squamous cell carcinoma and malignant melanoma, since the age of 5 till 22 years. Eventually the boy died at 22 years of age (Fig. 11.6). This is the sad part of this distressing and frustrating disease. I have not seen this girl since 12 years now

- Arsenic, once used in skin ointments, also increases the risk of basal cell carcinoma.

Clinical features

- The most common clinical presentation is an ulcer that never heals (Fig. 11.9). Sometimes healing takes place with scabbing and later it breaks down and forms ulcer again. The ulcer has ***a raised and beaded edge***, may be indurated and bleeds on touch. The base can be subcutaneous fat or deeper structures like muscle or bone depending upon invasion.
- When the ***lesion is big***, it is called ***rodent ulcer*** (Figs 11.10 and 11.11).

KEY BOX 11.1

BASAL CELL CARCINOMA: TYPES

- Nodular: Nodulocystic or noduloulcerative—70%
- Pigmented
- Superficial: Occur on the trunk
- Cystic
- Infiltrative
- Basosquamous: Rare

Fig. 11.8: Basal cell carcinoma—typical site

PEARLS OF WISDOM

Scabbing occurs only in benign ulcers. Basal cell carcinoma is the only malignant ulcer which shows scabbing.

- It can also present as a ***painless, firm, nodule,*** which is pigmented with fine blood vessels on its surface (Key Box 11.1).
- It can present as ***nodulo-ulcerative form*** (Fig. 11.8).

Fig. 11.9: Nonhealing ulcer of 3 years duration—lesion that does not heal and grows slowly—classical history of BCC

Fig. 11.10: Pigmented large destructive BCC (*Courtesy:* Dr Vidyadhar Kinhal, HOD, Surgery, VIMS, Bellary, Karnataka)

Fig. 11.11: Lateral view of the same patient showing elevated edge, few areas of scab and slough

- Rarely, it can be ***nodulocystic*** variety which does not show fluctuation.
- **Field fire rodent ulcer** is a rapidly growing rodent ulcer with destruction and disfigurement of the facial skin. It has an advancing edge with healed central scar.
- More aggressive infiltrative form is ***Morpheaform*** and ***more common*** form is ***nodulo-ulcerative form.***

Less aggressive: Nodular, superficial

Highly aggressive: Sclerosing (Morpheaform), infiltrating, micronodular, basosquamous

Differential diagnosis

- **Keratoacanthoma:** It occurs only in the face. The edge may be raised with ulceration, thus resembling basal cell carcinoma.
- **Sclerosing angioma**
- **Malignant melanoma:** Pigmented basal cell carcinoma may be mistaken for malignant melanoma.
- **Squamous cell carcinoma**

Spread

- It spreads by **local invasion**. Even though it is slow-growing, it slowly penetrates deep inside, destroys the underlying tissues such as bone, cartilage or even eyeball. Hence, the name rodent ulcer. It **does not spread by lymphatics** because the large size of tumour emboli. Blood spread is extremely rare.
- Morpheic BCC synthesises type IV collagenase and so spreads rapidly. White scar like growth pattern is seen in morphic variety.

KEY BOX 11.2

MICROSCOPIC PICTURE

- Central mass of polyhedral cells
- Cells are darkly stained
- With peripheral palisade layer of columnar cells. Cell nests, keratinisation and mitotic figures are absent.

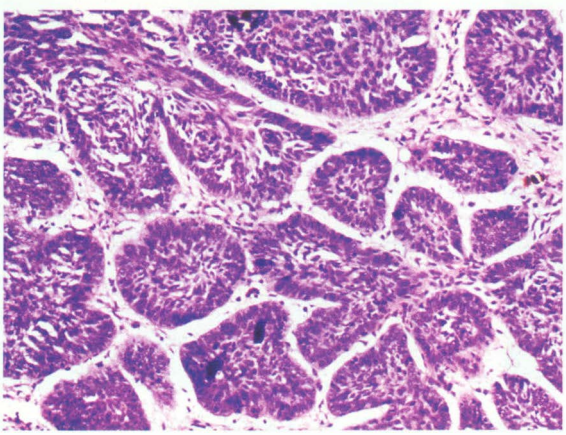


Fig. 11.12: Palisading islands of basaloid cells seen in BCC (*Courtesy*: Dr Laxmi Rao, HOD, Pathology, KMC, Manipal)

TNM staging

TNM STAGING	TNM Staging for skin cancer other than melanoma
T1	Tumour < 2 cm in greatest dimension with less than 2 high-risk features
T2	Tumour > 2 cm in greatest dimension (or) tumour of any size with 2 or more high-risk features.
T3	Tumour invasion of maxilla, mandible orbit or temporal bone.
T4	Tumour with invasion of skeleton or perineural invasion of skull base
N0	No regional lymph nodes
N1	Metastasis in a single ipsilateral lymph node ≤ 3 cm
N2	Metastasis in a single ipsilateral lymph node > 3 cm but < 6 cm; or B/L or contralateral lymph nodes none > 6 cm in greatest dimension
N3	Metastasis to lymph node > 6 cm in greatest dimension
M0	No metastases
M1	Distant metastases

Investigations

Wedge biopsy from the edge of the ulcer. The edge is selected because of the following reasons:

- Edge is the growing part, malignant cells are numerous
- Centre has slough or scab which may not reveal malignancy
- Comparison with normal skin is possible.

Histology (Key Box 11.2 and Fig. 11.12)

Types:

1. Basosquamous
2. Morpheaform
3. Adenoid
4. Infiltrative

Treatment (Fig. 11.17)

- Basal cell carcinoma responds well to radiation. Surgical excision also cures the disease.
- Surgery is the first line of treatment for basal cell carcinoma.

I. Surgery is indicated when the lesion is:

- Very close to the eye, adherent to the cartilage or bone
- In easily, accessible sites such as neck and hand.
- In radiation failure cases.

Wide excision is done in all cases: This means excision of the growth with at least 3–4 mm of healthy margin all around including at the base. The resulting defect is closed by:

a. **Primary suturing** of the defect if the lesion is small (Figs 11.18 to 11.20)
b. **Skin grafting** if defect is big as in neck or dorsum of hand
c. **Rotation flaps** as in face for better cosmetic effect (Figs 11.13 to 11.16).

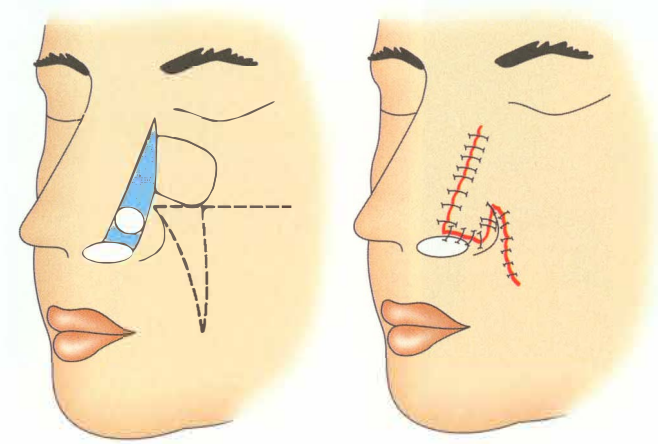


Fig. 11.13: Reconstruction with nasolabial flap following excision

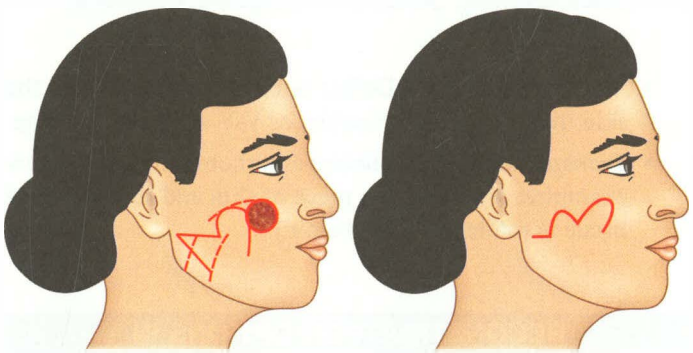
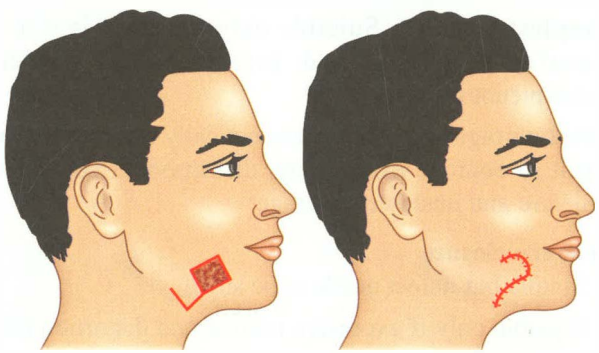
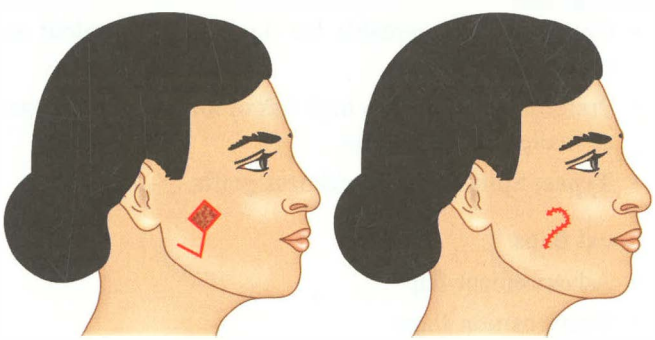


Fig. 11.14: BCC excision and reconstruction by using bilobed flap



Rhomboid flap



Banner's flap

Figs 11.15 and 11.16: BCC reconstruction

Mohs' micrographic surgery: It is a special surgical technique which involves excision of skin cancer under microscopic control. It minimises recurrence and maximises conservation of surrounding normal tissue.

The technique offers complete evaluation of the lateral and deep margins of the tumour excision.

Indications for Mohs' procedure

- Centrofacially located tumours
- Large tumours
- Poorly defined tumour margins
- Recurrent lesions
- Lesions with perineural/perivascular involvement
- Tumours at site of prior radiation therapy
- Tumours in the setting of immunosuppression
- High-risk histological subtype of BCC

Advantage

Better cosmetic results since minimal amount of normal tissue is removed.

II. Radiation is indicated in elderly patients who have an extensive lesion which requires a complicated plastic reconstruction. ***Dosage:*** 4000–6000 cGy units. Radiation chondritis and osteitis are the complications (*see* Fig. 11.17 for summary of the treatment of basal cell carcinoma).

III. Other forms of treatment for basal cell carcinoma

- Small and superficial: Curettage with electrodesiccation
- Liquid nitrogen for tumours less than 1 cm in diameter
- CO_2 laser in basal cell carcinoma (Key Box 11.3)

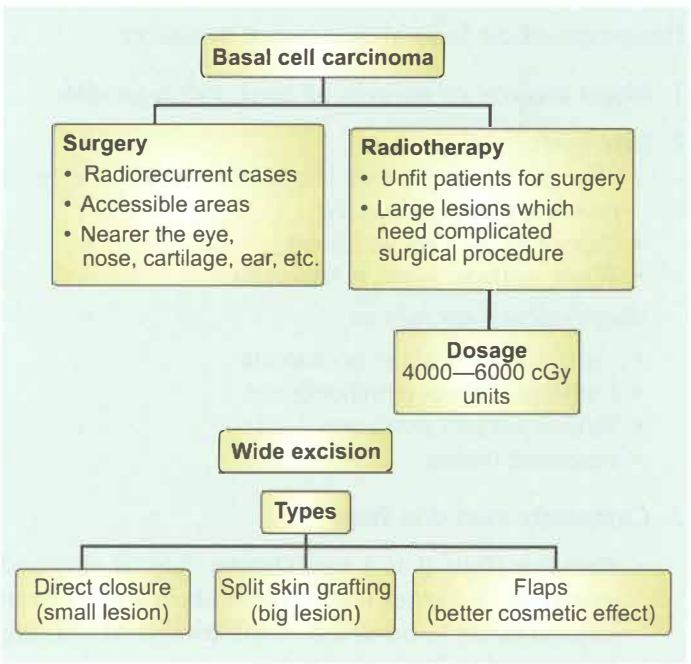


Fig. 11.17: Treatment of BCC

Fig. 11.18: Pigmented, elevated lesion—5 years duration

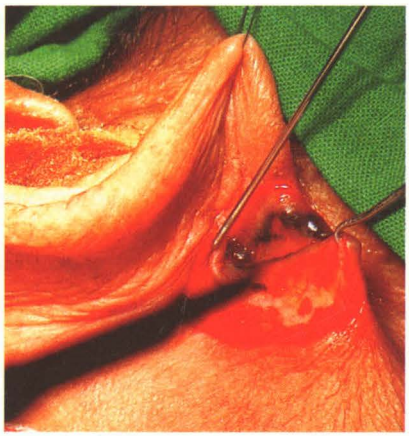


Fig. 11.19: Wide excision in progress

Fig. 11.20: Primary closure

(*Courtesy:* Dr Balakrishna Shetty, Professor of Surgery, KS Hegde Medical Academy, Deralakatte, Mangalore, Karnataka)

KEY BOX 11.3

CO_2 LASER IN BASAL CELL CARCINOMA

- Useful in superficial BCC that are confined to epidermis and papillary dermis.
- At least 4 mm surrounding healthy skin should be removed.
- Patients with multiple tumours or in hereditary Gorlin's syndrome—laser is very useful.
- Acts by superficial vaporisation
- Pathological margins cannot be examined.
- Indication in high risk BCC—large (> 2 cm), located near eye, nose, ear.
- Precancerous/low grade BCC

Reconstruction following tumour excision

1. Direct closure: *In majority of cases, this is possible.*

2. Skin graft

- Skin graft does not look like normal skin. However, it provides a good skin cover
- Cannot be used across a joint
- When aesthetic result is important.

Skin graft will not take on

- Cortical bone without periosteum
- Cartilage without perichondrium
- Tendon without paratenon
- Irradiated tissues.

3. Commonly used skin flaps

- ***Rotation flap:*** It is a semicircular flap of skin and subcutaneous tissues which rotates about a pivot point into the defect to be closed. Small triangle of skin can be removed to facilitate rotation.
- ***Advancement flap:*** Defect is closed by stretching the skin. It can be single, double, or V-Y advancement flap.
- ***Transposition flap:*** Bilobed and rhomboid flap are few examples. A rectangular piece of skin and subcutaneous tissue is rotated (details below).

RECONSTRUCTION OPTIONS IN HEAD AND NECK

1. Healing by secondary intention

Simplest method: Suitable only if defect is < 1 cm, superficial defects and located in cosmetically inconspicuous sites.

Disadvantage: Not always cosmetically acceptable, might result in contour irregularity, distortion of surrounding structure and unstable coverage.

2. Primary closure

- Simple but defect needs to be small.
- Possible only if excessive tension and distortion can be avoided.

3. Skin grafts

- Cosmetically acceptable but does not give colour and contour match.
- Preferred if patient has high risk of local recurrence such as malignant melanoma.
- **Types:** Split and full thickness grafts.

4. Local flaps

- Advancement flap
- Transposition flaps
- Rotation flaps, e.g. cervicopectoral flap

5. Local composite flaps

Flaps like PMMC flap, deltopectoral flap

6. Free flap

Radial artery based free forearm flap, fibular free flap

LOCAL FLAPS

Advancement flaps

- V-Y flap
- Good for defects on medial cheek and ala of nose.

A. V-shaped incision is made and then advanced to cover the defect and later sutured in a 'Y' shape.

B. Transposition flap

1. **Banner flap:** Provides excellent contour and reasonable colour match.
   - Can be used for small defects only.
   - **Disadvantage:** Produces a conspicuous scar.
2. **Bilobed flap:** Banner flap uses a single lobe. Instead, in this flap, two lobes are created and then rotated.
   - Used when defect is large enough not to be closed with banner flap.
   - Especially useful in area around the nose.
   - Produces conspicuous scar and is less desirable than banner flap.
3. **Rhomboid flap:** Geometric modification of Banner's flap. It can be used for medium-sized defects (details in page 149).

RECONSTRUCTION OPTIONS FOR LIPS

Most important factors that dictates reconstruction is amount of remaining lip vermilion.

A. Primary closure for small defects

B. Vermilion advancement flap

C. Abbe Flap

- Mainly used for upper lip
- It is harvested based on labial artery
- Provides good cosmetic effect
- Causes minimal loss of function in lower lip. Modification of Abbe flap like reverse Abbe flap can be used for lower lip defects also.

D. Estlander flap

- Rotates the upper lip to lower lip
- Disadvantage: Produces a round commissure and loss of normal taper of vermilion.

E. Webster-Bernard flap

- Medial advancement of cheek tissue to create a new lower lip.
- Gives a good result
- **Disadvantage:** Results in significant facial scarring.

Very large lower lip defects can also be closed with 'Double central severe Abbe's flap' or **Bilateral Karapandzic flap** (*see* page 289).

SQUAMOUS CELL CARCINOMA (EPITHELIOMA)

It is the **second common malignant skin tumour** after basal cell carcinoma. It arises from prickle cell layer of the Malpighian layer of the skin. It usually affects elderly males. All the premalignant conditions listed earlier apply to this condition. It can also occur *de novo* in the skin. Basosquamous carcinoma is the term applied when squamous cell carcinoma arises in a pre-existing basal cell carcinoma. It is interesting to note that a variety of names have been given to squamous cell carcinoma when it occurs in different places. They are given in Table 11.2.

Pathological types

- Ulcerative variety—commonest
- Cauliflower like or proliferative growth
- Ulceroproliferative type

See Table 11.3 for the typical sites of skin, mucous membrane and junction involvement.

Table 11.2 Sites of squamous cell carcinoma

Site	Name	Reason/explanation
Skin of the abdomen or back of thigh in Kashmiri patients	**Kangri cancer**	**Kangri** is the name given to the **pot containing hot charcoal** which is applied to the abdominal wall because of excessive cold in Kashmir (India)
Buttocks, heels, elbows	**Kang cancer**	Tibetans sleep on the **oven bed** due to excess cold
Scrotum	**Chimney sweep cancer**	Seen in **chimney sweepers** due to prolonged irritation by chemicals such as tar or pitch in
Abdominal wall	***Saree* cancer and *Dhothi* cancer**	Due to chronic irritation caused by wearing *dhothi* or *saree* too tight
Lower lip	**Countryman's lip**	Carcinoma lower lip is common in **agriculturists** (outdoor occupation)

Table 11.3 Typical sites of squamous cell carcinoma

Skin	Junction	Mucous membrane
• Dorsum of hand • Limbs • Face • Abdominal wall	• Between skin and mucous membrane • Lip • Penis • Vulva	• Lined by stratified squamous epithelium as in oral cavity and oesophagus • Lined by columnar epithelium, wherein squamous metaplasia occurs, such as gall bladder, bronchus • Lined by transitional cell epithelium with metaplasia as in urinary bladder

Clinical features

- Most lesions are preceded by actinic keratosis. Such lesions grow slowly and are locally invasive without metastasis. If not from keratosis, lesions are more aggressive and metastatic.
- Typically, it is an ulcerative or cauliflower-like lesion (Fig. 11.21).
- The edge is everted and indurated (Fig. 11.22).
- The base is indurated and it may be subcutaneous tissue, muscle or bone.
- The floor contains **cancerous tissue which looks like granulation tissue.** It is pale, friable, bleeds easily on touch (Figs 11.23 to 11.27).
- Surrounding area is also indurated.
- Mobility is usually restricted due to infiltration of underlying structures. In very early cases, ulcer can be moved along with skin over the underlying structures.
- Regional lymph nodes such as inguinal lymph nodes (both vertical and horizontal groups) can get enlarged when squamous cell carcinoma affects lower limb or abdominal wall. Hard lymph nodes are suggestive of secondaries.

Spread

1. **Local spread** occurs by infiltration into the surrounding tissues. Depending upon the site, various structures can be involved. Some examples are:
   - Tendon involvement in the dorsum of the hand.
   - Muscle involvement in the abdominal wall.
   - Bone involvement, e.g. tibia in carcinoma developing in a varicose ulcer or mandible in carcinoma cheek.
2. **Lymphatic spread** is the chief method of spread even though it occurs relatively late. Regional nodes are involved first.
   - Nodes which are soft to firm and tender are due to secondary infection.
   - Nodes which are hard, nontender, with or without fixity are due to secondary deposits.
   - In untreated cases, nodes start ulcerating through the skin resulting in bleeding and pain.
   - As already stated, nodes do not get involved in Marjolin ulcer.
3. **Blood spread** is rare and late.

PEARLS OF WISDOM

Epithelial pearls are absent in poorly differentiated carcinomas.

Differential diagnosis (Key Box 11.4)

Investigations

- A wedge biopsy from the edge of the ulcer or growth is taken. However, in proliferative lesions punch biopsy can also be taken.
- **Microscopic picture:** Eighty per cent of these cancers are well differentiated (Key Box 11.5). It is characterised by central structureless mass of keratin surrounded by normal looking squamous cells which are arranged in **concentric manner like onion skin.** This whole appearance is called **epithelial pearl or cell nest** (Fig. 11.28). In 20% of cases, cells are **undifferentiated** with numerous mitoses, without keratinisation.

KEY BOX 11.4

DIFFERENTIAL DIAGNOSIS

• Basal cell carcinoma	: Typical sites
• Keratoacanthoma	: Benign self-limiting
• Papilloma	: Benign lesion
• Pyogenic granuloma	: Painful, short duration
• Tuberculous ulcer	: Uncommon

KEY BOX 11.5

BRODER'S CLASSIFICATION

I. Well differentiated	75% keratin pearls
II. Moderately differentiated	50% keratin pearls
III. Poorly differentiated	25% keratin pearls
IV. Anaplastic	< 25% keratin pearls

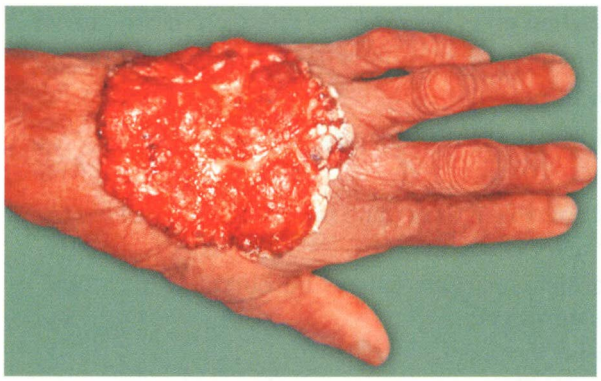


Fig. 11.21: Squamous cell carcinoma affecting dorsum of the hand—ulceroproliferative lesion

Fig. 11.22: Squamous cell carcinoma—bleeds on touch

Fig. 11.23: Epithelioma scalp—large ulcerating bleeding lesion

Fig. 11.24: Squamous cell carcinoma arising in leprosy scar

Fig. 11.25: Squamous cell carcinoma of the sole

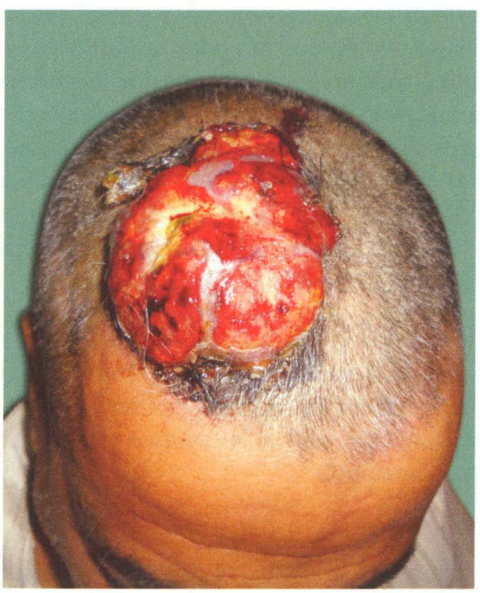


Fig. 11.26: A large epithelioma of the scalp—biopsy proved squamous cell carcinoma

Fig. 11.27: Bowen's disease with squamous cell carcinoma

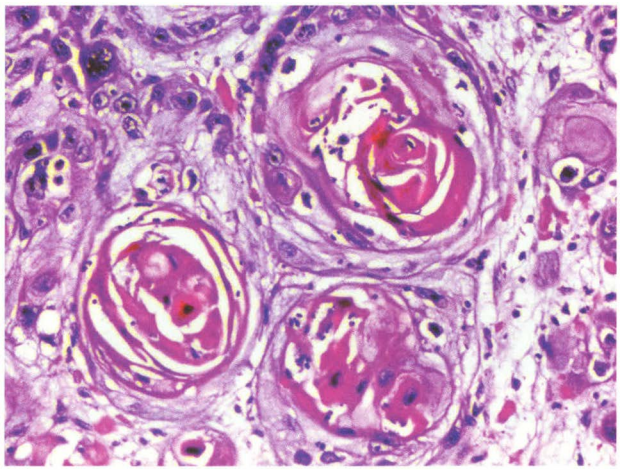


Fig. 11.28: Classical keratin pearls—well-differentiated squamous cell carcinoma (*Courtesy:* Dr. Laxmi Rao, Head, Department of Pathology, KMC, Manipal)

Treatment

Treatment can be classified as treatment of the primary and treatment of the secondaries.

I. Treatment of the primary

Squamous cell carcinoma is treated by wide excision or radiotherapy.

A. Surgery (Figs 11.29 and 11.30)

- It involves removal of growth along with 1 cm of normal healthy tissue from the palpable indurated edge of the tumour, in high-risk cases.
- Low-risk lesions can be excised with 4–6 mm margin.

Indications for surgery

- When the lesion is small and superficial
- When the lesion has involved deeper tissues such as muscles, cartilage or bone
- Radiorecurrent cases

Reconstruction: After wide excision, the defect can be closed primarily, with split skin graft or with a flap to reconstruct the part depending upon the extent of resection. If the growth i fixed to tibia, below knee amputation is the treatment.

Fig. 11.29: Wide excision of squamous cell carcinoma

Fig. 11.30: Wide excision followed by skin grafting on 3rd year follow-up

B. Radiotherapy

Indications for radiotherapy

Primary

1. Patients unable or unwilling to undergo surgical treatmen for primary lesion
2. When clear margins cannot be obtained by Mohs' o extensive surgery
3. Adjuvant treatment
4. Histologically aggressive, e.g. perineural invasion
5. High grade
6. Bone invasion (Fig. 11.31)
7. Lymph nodes—adjuvant or palliative intent

II. Treatment of secondaries

1. Thirty to forty per cent of the enlarged regional nodes are due to secondary infection. Once the primary is treated and antibiotics are given, lymph nodes may regress. In such cases, 'wait and watch policy' is observed.
2. If lymph nodes do not regress or are hard and mobile, FNAC can be done to confirm the diagnosis followed by radical block dissection. Thus, squamous cell carcinoma of the leg requires inguinal block dissection.
3. If lymph nodes are hard and fixed to the femoral vessels, palliative radiotherapy is given. Even in advanced fungating lesions, the response rate to radiotherapy is reasonably good.

- Dose: 3000–4000 cGy units over 3–4 weeks, 200 units/day.

Structures removed in inguinal block dissection

- The superficial group of nodes consisting of a horizontal chain which lies below inguinal ligament and a vertical chain which lies along upper 5–6 cm of long saphenous vein. These two groups of nodes form the letter T.
- The deep glands are located alongside the proximal end of the femoral vein, and one lying within femoral canal.
- Fat, fascia, lymphatics are cleared from 2 cm above the inguinal ligament up to 2 cm below saphenofemoral junction. The medial clearance is important up to femoral canal. 8–10 cm long saphenous vein near its termination is removed to facilitate lymph node clearance.

Inguinal block dissection

- The saphenous vein can be preserved, nodal tissue is removed from the vessel circumferentially; otherwise, it is sacrificed.
- Cloquet's node is sent as a separate specimen for frozen section examination

PEARLS OF WISDOM

Fixity to muscle, veins, fascia and bones is not a contraindication for block dissection. Carotid and femoral artery infiltration makes it inoperable.

Fig. 11.31: Large penetrating baso-squamous cell carcinoma with destruction of the left ear and infiltrating mastoid process and external ear

Complications of inguinal block dissection and treatment

- Wound infection → Broad-spectrum antibiotics
- Lymphorrhoea → Adequate drainage
- Haemorrhage → Perfect haemostasis
- Flap necrosis → Avoid sharp corner
- Femoral blow-out → Sartorius muscle slide to cover femoral vessels at the end of surgery

Summary of the treatment of squamous cell carcinoma (Fig. 11.32)

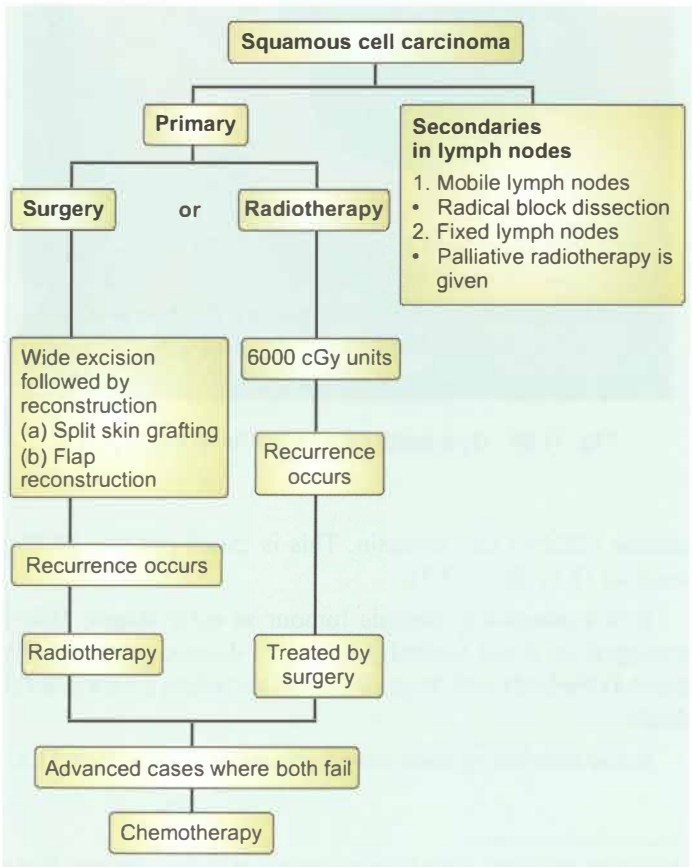


Fig. 11.32: Summary of the treatment of squamous cell carcinoma

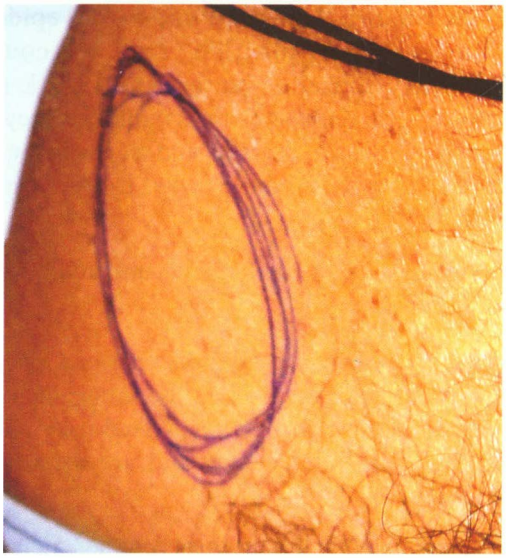


Fig. 11.33: Hard lymph node in the deep inguinal region suggestive of metastasis—see clinical notes

Prognostic factors (Key Box 11.6)

KEY BOX 11.6

PROGNOSTIC FACTORS OF SCC

- *Invasion:* Deeper the lesions or SCC less than 2 mm, metastasis is unlikely. If it is greater than 6 mm—15% SCC may have metastasised
- *Broder's grade higher:* Worse prognosis
- *Site:* Lip and ear—high chances of recurrence
- *Immunosuppression:* Poor prognosis
- *Perineural involvement:* Worse prognosis

CLINICAL NOTES

A patient who had undergone wide excision and SSG for a growth behind the knee, presented with oedema of the leg. The MBBS student could not explain oedema. He said the cause is nutritional. Groin had 3–4 hard nodes.

MELANOCYTIC TUMOURS

Simple melanocytic tumours

These are also called pigmented naevi which are composed of modified melanocytes derived from the neural crest. All naevi have excess melanin pigmentation because of which they are tan brown or black in colour. They are located in the basal layer of the epidermis. They are benign. They are of following types:

1. **Junctional naevus:** Located within the epidermis at the dermoepidermal junction. They are common in children. They appear as tan brown to black macules. They are smooth, flat and hairless moles. As they enlarge, they become slightly raised and can evolve into an intradermal or a compound naevus. It has no malignant potential.

   Junctional naevus commonly occurs on the palm, sole, digits and genitalia.

2. **Compound naevus:** As the mole enlarges, naeval cells also appear in the dermis along the intraepidermal cells. Such moles are described as compound naevi. These are found usually in adolescents and are usually benign (Fig. 11.34).

3. **Intradermal naevus:** It is the most common mole in adults. Because of its deep seated nature, it appears blue. Hence, the name blue naevus. It is seen on the scalp and face. It contains hair and does not become malignant.

4. **Spitz naevus:** Commonly occur on face and legs. Grows rapidly initially and then remains static. It appears as reddish brown nodule—occasionally deeply pigmented.

5. **Spindle cell naevus:** Occur in women. Common site is the thigh. It has a malignant potential.

6. **Congenital pigmented naevus** which is present at birth, has a **greater potential for malignant change**. It can involve extensive areas of the skin **(Giant)**.

   Giant congenital pigmented naevus (GCPN) or giant hairy naevus.

   - It is an example of hamartoma of naevomelanocytes. Involves extensive areas of the skin.
   - Naevus cells are present not only in epidermis but also in the subdermal fat and muscle
   - Risk of malignant melanoma is about 10%
   - Malignant melanoma tends to be axial
   - Removal is for aesthetic and oncological reasons.

7. **Dysplastic naevi** are different from acquired naevi in the following ways:
   - Malignant potential is more (Figs 11.35 and 11.36)
   - Family members may have such lesions.
   - Such syndrome is described as **familial dysplastic naevus syndrome**.

Fig. 11.34: Compound naevus

Fig. 11.35: Dysplastic naevi over the face

Fig. 11.36: Dysplastic naevus left temporal region

MALIGNANT MELANOMA[1] (MELANOCARCINOMA)

It is a malignant tumour arising from pigment-forming cells (melanoblasts) which are derived from the neural crest. Melanoblasts and melanocytes convert dihydroxyphenylalanine (DOPA) into melanin. This is called positive DOPA reaction (Key Box 11.7).

It is a potentially curable tumour in early stages. If left untreated or if not treated properly, it disseminates rapidly, showers the body with tumour emboli and offers a very painful death.

Some interesting most have been given in Key Box 11.8.

[1]Malignant melanoma is the killer skin cancer of whites. "Bronzed Body Beautiful" concept should be discouraged.

KEY BOX 11.7

POSITIVE DOPA REACTION

TYROSINE
↓ ← TYROSINASE
DOPA
↓ ← OXIDASE
MELANIN

KEY BOX 11.8

INTERESTING 'MOST' FOR MALIGNANT MELANOMA

- Most benign form of melanoma is lentigomaligna melanoma.
- Most common form of melanoma is superficial spreading melanoma.
- Most malignant type of melanoma is nodular melanoma.
- Most reliable independent prognostic indicator in malignant melanoma is tumour thickness based on Breslow classification.
- Most of them (majority) arise from pre-existing moles.
- Most of the melanoma are pigmented. However, pigmentation is not mandatory for diagnosis of melanoma.

Common sites of malignant melanoma

- Head and neck 20–30%
- Lower extremity 20–30%
- Trunk 20–30%
- Remaining cases occur in upper extremities, genitalia, choroid of the eye[1].

Aetiopathogenesis (Fig. 11.37)

For risk factor study (Key Box 11.9)

1. **Ultraviolet rays:** It is more common in white-skinned people. There is a linear correlation between intensity of exposure to sunlight and malignant melanoma in white-skinned people[2]. White-skinned people who live close to the equator have increased tendency of developing malignant melanoma. The highest incidence is found in Queensland (Australia). For the same reasons, malignant melanoma is common in the United Kingdom, North America and Australia.

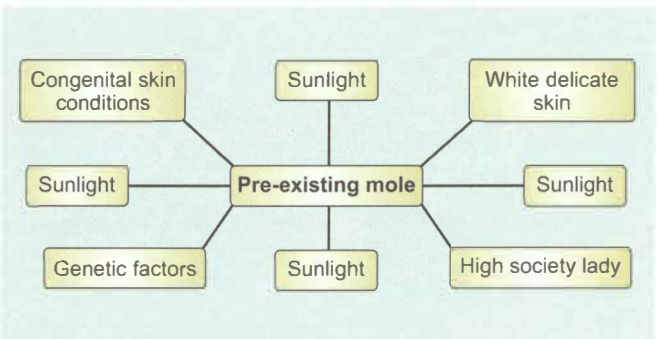


Fig. 11.37: Aetiopathogenesis

2. **Age and sex:** Malignant melanoma is more common in females. The higher incidence of the disease is found during reproductive age period. Even though oestrogen and progesterone receptors are found in malignant melanoma in some patients, their true role is not yet established. Leg is the commonest site in females.

   In general, the incidence of melanoma is higher in men than in women. The incidence of melanoma is 1.7 times higher for women than men before 39 years of age. After 70 years of age, the incidence of melanoma is 2.2 fold higher for men than women.

3. **Genetic factors**
   - Tumour suppressor gene mutation 9p21
   - Deletion or rearrangement of chromosomes 10 and 8p.
   - Familial atypical multiple, mole, melanoma syndrome—previously known as dysplastic naevus syndrome
   - Multiple, large > 5 cm, atypical dysplastic nevus in areas covered by clothing.
4. **Genetic factors:** Increased incidence has been found in patients with familial dysplastic naevus syndrome. The disease is also common in individuals with Celtic race who give family history of malignant melanoma (3–5%).
5. **Trauma:** Malignant melanoma occurs in the sole of the foot in African blacks. Whether trauma is the cause is not clear.
6. **Pre-existing mole:** Approximately 1/3rd of melanomas arise in a pre-existing mole, remaining 1/3rd arise *de novo* in the normal skin. Malignant change occurs in the junctional or compound naevus. Malignancy should be suspected when following changes occur in a mole (Key Box 11.10).

KEY BOX 11.9

RISK FACTORS FOR DEVELOPING MALIGNANT MELANOMA (MM)

- **P**igmentosa xeroderma
- **I**mmunocompromised—HIV, Hodgkin's disease treated with cyclosporin
- **G**enetic—History of dysplastic naevus
- **M**elanoma excised earlier
- **E**arly childhood burns and eyes blue, red hair
- **N**aevus: GCPN and total number > 20 naevi
- **T**endency to freckle

Remember as **PIGMENT**

[1]Choroid has no lymphatics. Hence, excellent prognosis.
[2]Malignant melanoma is the killer skin cancer of whites. "Bronzed Body Beautiful" concept should be discouraged.

KEY BOX 11.10

GLASGOW—CHECK LIST (Figs 11.38 and 11.39)

1. Change in size
2. Change in shape
3. Change in colour
4. Inflammation
5. Crusting, ulceration, bleeding
6. Sensory change
7. Diameter > 6 mm

7. **Increased incidence** of malignant melanoma has been found in patients with renal transplantation and leukaemia as a result of immunosuppression.
8. **Xeroderma pigmentosum and albinism** patients are susceptible for melanomas.

PEARLS OF WISDOM

"Bronzed, Blue Eyed, Blond, Body Beautiful—high chances of malignant melanoma"

Pathology

Microscopic picture: Anaplastic, pigment laden melanocytes confined to epidermis. The cells which have vacuolated cytoplasm (Paget's cells resembling those seen in Paget's disease of the breast) are found. Cells also invade the dermis. Along with pigment-laden macrophages, dermal infiltration of lymphocytes[1] may be present. Rarely, anaplastic melanocytes do not form pigment **(amelanotic melanoma)**.

All melanomas (except nodular) show **radial growth initially** in the form of intraepidermal growth. However, **nodular melanoma** has **vertical growth phase** thus involving dermis leading to nodule formation. This has poor prognosis.

Pathological grading of malignant melanoma
(Fig. 11.40)

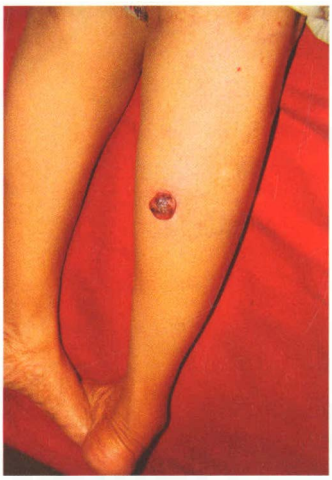
Clarke's level of invasion

Level 1	Tumour cells confined to basement membrane
Level 2	Tumour extension into papillary dermis
Level 3	Tumour reaches the interface between papillary dermis and reticular dermis
Level 4	Tumour reaches reticular dermis
Level 5	Tumour invades subcutaneous fat.

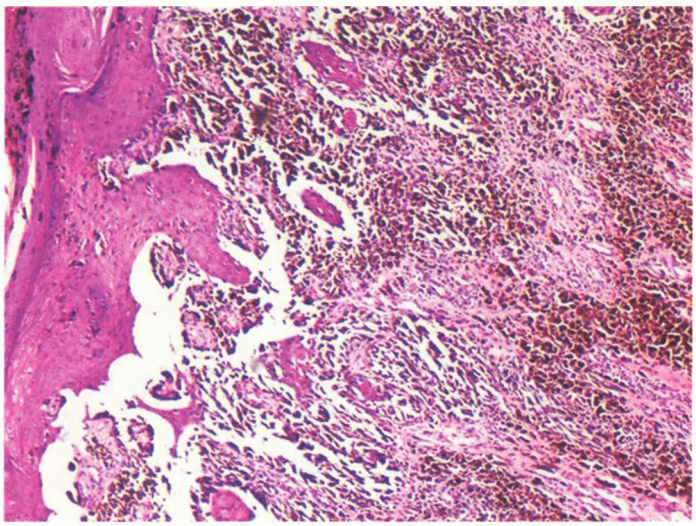
It is no longer followed.

Breslow described staging depending upon the maximum thickness at the centre of the lesion.

[1]Presence of lymphocytes may be an indication of host response 'Fight' against cancer.



Figs 11.38 and 11.39: Only complaint of this 28-year-old lady was an increase in the size and oozing from the lesion



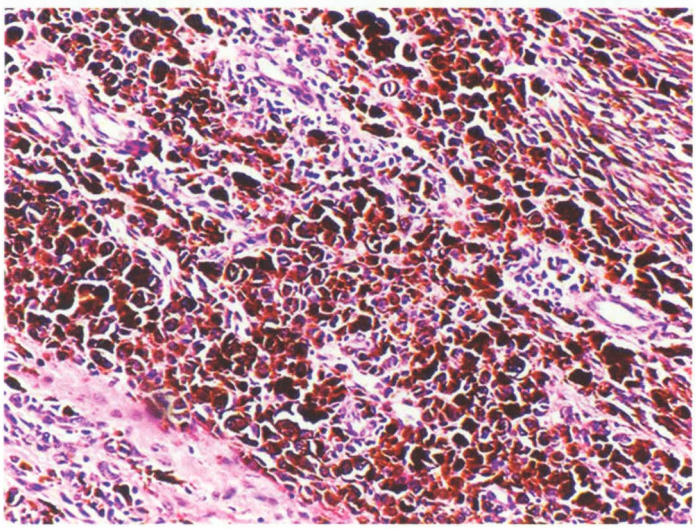


Fig. 11.40: Malignant melanoma with malignant cells showing dark melanin pigment inside. Second picture is a close-up view (*Courtesy:* Dr Laxmi Rao, HOD, Pathology, KMC, Manipal)

Table 11.4 Clinical types of malignant melanoma (Figs 11.41 to 11.44)

Name	Nature	Behaviour
Lentigo malignant melanoma	Arises from Hutchinson's melanotic freckle Occurs in old people on face and temporal region	Least common (5%) Least malignant
Superficial spreading	Any part, more in the trunk	Most common (70%)
Nodular	Any part, more in the leg	Most malignant, invasive (15–30%)
Acral lentiginous	Located on the palm, sole, digits	Presents late, aggressive (4–8%)
Amelanotic	Difficult to diagnose	Rare, nonpigmented (5%)

Stage I : Thickness less than 0.75 mm
Stage II : 0.76 to 1.5 mm
Stage III : 1.51 to 3.0 mm
Stage IV : More than 3 mm

- **Less than 1 mm is regarded as thin melanoma.**

Clinical type (Table 11.4)

Clinical features

- Malignant melanoma can present as changes in the pre-existing mole which are already described (Key Box 11.11).
- The patient can present as a nonhealing ulcer of the sole of the foot.
  - It is a painless ulcer
  - Edges are irregular
  - Floor is irregular
  - Bleeds on touch
  - Typically, the **ulcer is pigmented** (Fig. 11.41). In 10% of patients, pigment is absent. They are called amelanotic melanoma.
  - Lesion is firm in consistency and **induration is absent**.
  - A **halo** may be present surrounding the ulcer.
  - The lesion moves with the skin and is usually not fixed to underlying structures.
  - **Satellite nodules** (within 5 cm of the primary) may be found surrounding the lesion which are due to spread through intradermal lymphatics (Fig. 11.47). Such patients will have greatly enlarged, firm, nontender nodes.
  - **In-transit lesion**—disease found in the dermis or subcutaneous tissue more than 2 cm away from the primary melanoma but before the regional lymph node basin (Figs 11.45, 11.46 and 11.48)
    See Key Box 11.12 for acral lentiginous variety.

KEY BOX 11.11

ABCDE OF EVALUATING A CHANGING MOLE

A Asymmetry: One half does not match the other
B Border irregular: Ragged or blurred
C Colour variation: Tan, black, brown
D Diameter: > 6 mm
E Evolving (elevation): Change in a pre-existing lesion

KEY BOX 11.12

ACRAL LENTIGINOUS (Fig. 11.43)

- Least common subtype and very aggressive
- Popularly called hand and foot melanoma
- Occurs in palms and soles
- This subtype occurs in dark-skinned people (21–22%).
- In subungual region—blue black discolouration of posterior nail fold (most common on great toe or thumb).
- Pigmentation in proximal or lateral nail folds (Hutchinson's sign)

AMELANOTIC MELANOMA (Fig. 11.44)

- Lesion appears pink
- It has poor prognosis than nodular melanoma.

PEARLS OF WISDOM

In all cases of suspected melanomatous skin lesions examine axillae, scalp, soles, genitalia, oral cavity and interdigital webs also.

TNM staging

TNM STAGING AJCC 6th Edition

Tumour
T0 No tumour
Tis *in situ* tumour
T 1a < 1 mm, level II, level III no ulceration
T 1b < 1 mm, level IV with ulceration
T2a 1–2 mm no ulceration
T2b 1–2 mm with ulceration
T3a 2–4 mm No ulceration
T3b 2–4 mm with ulceration
T4a > 4 mm no ulceration
T4b > 4 mm with ulceration

Node
N0—no nodes
N1a—one node micrometastasis
N1b—two nodes macrometastasis
N2a—2 or 3 nodes micrometastasis
N2b—2 or 3 nodes macrometastasis
N2c—no nodes but satellite or in-transit lesions
N3–4 more nodes; nodes with satellite or in-transits, matted nodes

M—Metastasis
M0—no blood spread
M1a—skin, subcutaneous tissue, distant node—normal LDH
M1b—lung spread—normal LDH
M1c—other viscera or distant spread and increase in LDH with any metastasis

Fig. 11.41: Superficial spreading

Fig. 11.42: Nodular

Fig. 11.43: Acral lentiginous variety with early changes—never incise or cauterisesuch melanomas

Fig. 11.44: The lesion resembles squamous cell carcinoma. Punch biopsy reportec as amelanotic melanoma

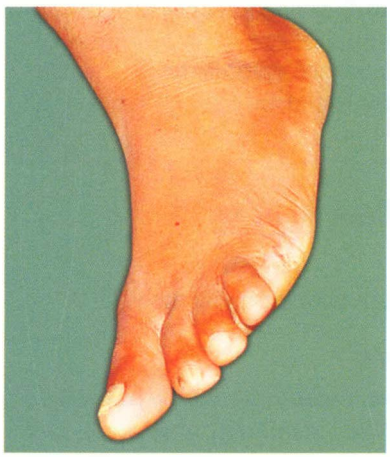


Fig. 11.45: Observe the foot, little toe has been amputated five years back for malignant melanoma

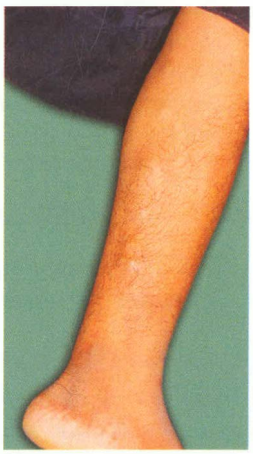


Fig. 11.46: Same patient as in Fig. 11.45 presented to the hospital with in-transit deposits

Fig. 11.47: Malignant melanoma operated—recurrence after 8 months with satellite nodule. It is important that the first surgery should be the best surgery. Every attempt should be made to do a three-dimensional excision

CLINICAL STAGING*			
Stage 0	Tis	N0	M0
Stage IA	T1a	N0	M0
Stage IB	T1b	N0	M0
	T2a	N0	M0
Stage IIA	T2b	N0	M0
	T3a	N0	M0
Stage IIB	T3b	N0	M0
	T4a	N0	M0
Stage IIC	T4b	N0	M0
Stage III	Any T	≥ N1	M0
Stage IV	Any T	Any N	M1

*Clinical staging includes microstaging of the primary melanoma and clinical/radiologic evaluation for metastases. By convention, it should be used after complete excision of the primary melanoma with clinical assessment for regional and distant metastases

Fig. 11.48: Whole leg is studded with tumour. In-transit deposits from malignant melanoma of the foot

Important changes in the new AJCC staging of malignant melanoma

1. Thickness and ulceration in T-category rather than level invasion
2. Number of metastatic nodes is more important
3. LDH is included in 'M' category (metastatic spread)
4. If there is ulceration, 3 stages are upgraded.
5. Satellite nodules and in-transit deposits are grouped into stage III disease.

Differential diagnosis

- Pigmented basal cell carcinoma
- Histiocytoma (sclerosing angioma)
- Naevus, Kaposi's sarcoma, cavernous haemangioma.

Spread

1. **Local spread** occurs mainly by continuity and contiguity. **Satellite nodules** are due to local and lymphatic spread, situated within 5 cm of the primary lesion. Malignant melanoma rarely infiltrates the deep fascia unless and until a 'blunder biopsy' is done. **Inadequate local excision can result in local recurrence later. Hence, initial surgery should attempt at cure.**
2. **Lymphatic spread** is the principal mode of spread. Regional nodes get involved very early in malignant melanoma, altering the prognosis. Thus, nodes can get enlarged to a large extent even when the lesion looks innocent. Spread occurs both by permeation and embolisation. Permeation produces satellite nodules and in-transit nodules which develop between primary and secondaries. Embolisation occurs rapidly and early producing massive regional nodes (Key Boxes 11.13 and 11.14).

KEY BOX 11.13

MALIGNANT MELANOMA SOLE

Inguinal nodes
|
Iliac nodes
|
Para-aortic nodes
|
Mediastinal nodes
|
Supraclavicular nodes

KEY BOX 11.14

LYMPH NODE METASTASIS IN MALIGNANT MELANOMA

- It is the single-most important prognostic index and it presents stage III disease.
- **Number of nodes is more important** than size of nodes.
- **Poor survival in ulcerated** malignant melanoma, even if node-negative.
- **Extranodal extension** has poor prognosis.
- Lymph node metastasis proceeds as an orderly process. Thus, evolved the concept of the **first node to get involved—sentinel node (SN).**
- **Elective lymphadenectomy** is **not indicated in thin melanoma** (1 mm)
- In **thick melanoma**, do a sentinel node biopsy. If positive, proceed to complete lymphadenectomy (block dissection).

In-transit metastasis appear in the skin as intracutaneous metastasis. They are thought to be due to melanoma cells trapped in lymphatic vessels.

3. **Blood spread** occurs relatively early and it causes secondaries in liver, lung, brain and bones producing miserable, pathetic situations. They are summarised in Table 11.5.
See the interesting clinical notes.

CLINICAL NOTES

A 55-year-old lady presented to the hospital with enlarged inguinal lymph nodes. Clinical examination of the leg revealed nodules in the leg with firm to hard enlarged nodes in the inguinal region. On careful observation, the little toe was missing. On questioning, the patient admitted that the toe had been amputated elsewhere five years back for a painless blackish lesion (Figs 11.45 and 11.46).

Investigations

- There is no specific investigation except an **excision biopsy** of the lesion. Excision with 1 cm of the margin is all that is required. Incisional biopsy is avoided because of the following reasons:

Table 11.5 Spread of malignant melanoma

Spread	Diagnosis	Problems
Metastasis in lung	Cannonball appearance, pleural effusion	Respiratory failure
Metastasis in liver	Massive hepatomegaly, ascites	Abdominal discomfort
Metastasis in brain	Raised intracranial tension	Coma
Metastasis in bone	Bony pains, pathological fractures	Paraplegia, quadriplegia
Metastasis in bowel	Bleeding	Anaemia

- It may injure the deep fascia and it may open up a new plane of spread.
- It does not allow the pathologist to perform a detailed histological examination (done in large lesions).

- **Nonspecific investigations** to look for metastasis are:
  - Chest X-ray—cannonball secondary
  - CT scan is better, however, it is costly (Fig. 11.49)
  - Ultrasound abdomen—secondaries in liver
  - X-ray of involved bone—osteolytic lesions
- **FNAC** of the regional lymph nodes
  - Fine needle aspiration of groin nodes is helpful in detecting the spread and to stage the disease (Figs 11.50 and 11.51).
- A large ulcerated lesion can be subjected to histopathology by punch biopsy or even incision biopsy.
- Serum LDH levels increase indicate metastatic disease.

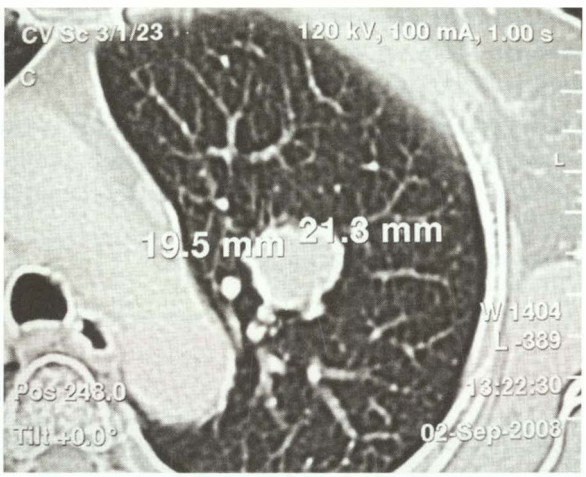


Fig. 11.49: Lung metastasis in CT of the chest. It was not visible in X-ray chest

Fig. 11.50: Hugely enlarged inguinal lymph nodes—the nodes can be firm to hard. Sometimes, they are soft due to degeneration

Fig. 11.51: FNAC of the recurrent inguinal node secondaries—bluish black aspirate

- HHB-4S-premelanosomal protein is specific immunohistochemical marker for melanoma.

Treatment

PEARLS OF WISDOM

Surgery is the main modality of treatment available for malignant melanoma. All other modalities of treatment are only palliative and supportive.

Types of surgery possible are as follows

1. **Excision biopsy-wide excision:** A small lesion of 1 mm can be excised even under local anaesthesia with 1 cm of healthy margin around (narrow excision). Defect can be closed by primary suturing. While excising the tumour, it is better not to handle the tumour. It is possible to remove the tumour by strictly adhering to the principle of 'No touch' technique (Figs 11.52 and 11.53). Wide excision is based on principle—centrifugal spread of melanoma.

Tumour thickness (mm)	*Wide excision margin*
< 1 mm	1 cm
1–2 mm	2 cm
> 2 mm	2 cm

   - Desmoplastic melanoma: Has high rate of recurrence, hence wide margin of excision is required.
2. **Clarke's level II lesions** are managed by a wider excision along with 2 cm of healthy margin around. Resulting defect is closed by split skin grafting (Fig. 11.54).
3. **Subungual malignant melanoma** is treated by amputation of the digit.
4. **Malignant melanoma of the choroid** has good prognosis. Choroid does not have lymphatic drainage. However, metastasis can occur through haematogenous route even after many years. It is treated by enucleation of the eye.
5. **Amputation (advanced and large lesions)** (Fig. 11.55).

Fig. 11.52: Ulcerated melanoma over the heel—poor prognosis

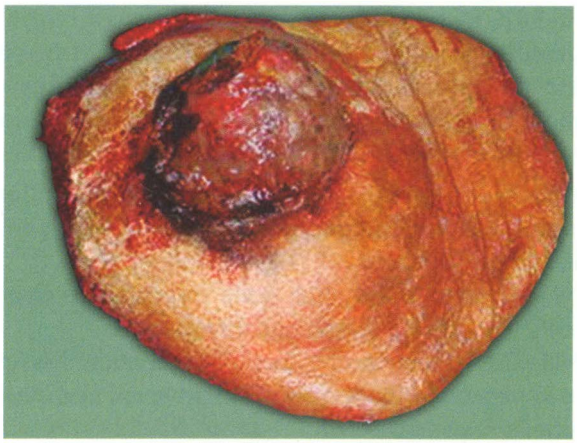


Fig. 11.53: Wide excision (minimum 2 cm) specimen

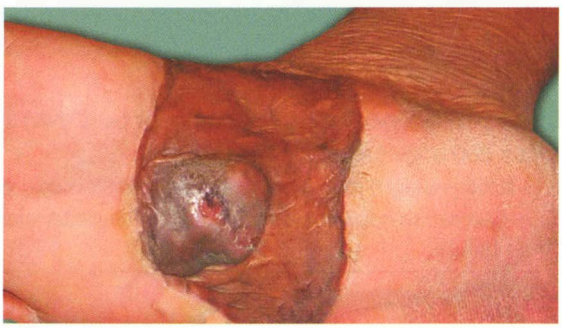


Fig. 11.54: Local recurrence after 2 years probably because of inadequate local excision—you can see the previous skin graft

Fig. 11.55: 8 cm ulcerated pigmented lesion in the foot—malignant melanoma foot required amputation

Fig. 11.56: Lazy S incision is given for inguinal block dissection—this incision decreases incidence of flap necrosis

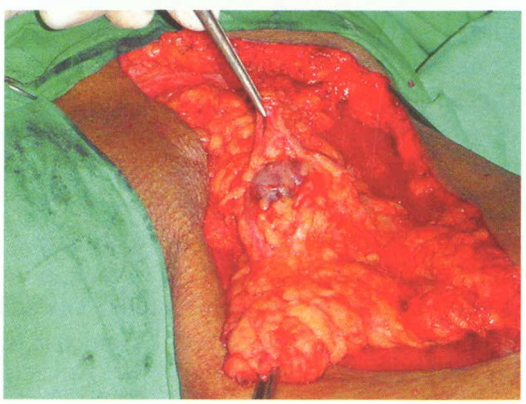


Fig. 11.57: Observe pigmented lymph nodes

Management of lymph nodes

1. If lymph nodes are situated adjacent to the primary lesion, block dissection is done along with primary lesion in continuity so as to include 'in-transit' deposits also.
2. If lymph nodes are away, radical block dissection is done. Example: For a lower limb malignant melanoma, inguinal nodes along with iliac nodes are removed. This is called **Ilioinguinal block** dissection. If these group of nodes are positive at frozen section, lymph node clearance should include lymph nodes of obturator vessels. This is called **Ilio-obturator block dissection** (Figs 11.56 and 11.57).

- **Sentinel lymph node mapping:** Isosulfan blue is injected intradermally and the node that gets stained is identified and sent for frozen section (Haematoxylin and Eosin stains and immunohistochemical technique). **If the node is positive, regional lymphadenectomy is done even when nodes are clinically not palpable**. At least

20–30% of the patients who are in Stage I will be identified as Stage III after sentinel node biopsy. There is a **definite survival advantage in patients** who undergo sentinel node biopsy (SNB) and regional lymphadenectomy when the nodes are not palpable.

- If SNB is negative, prophylactic block dissection is indicated only if melanoma has poor prognostic histological factors.

TLND—Therapeutic lymph node dissection is done in patients with known lymph nodal metastasis.

ELND—Elective or prophylactic lymph node dissection done in patients with no clinical lymph nodal metastasis is beneficial in patients with

- 1–4 mm thick melanoma
- Melanoma without ulceration
- Extremity melanoma
- Patients younger than 6 years of age

- Only 25% of patients slow occult metastasis of age

3. If lymph nodes are enlarged, hard and fixed, palliative radiotherapy is given.

Management of advanced malignant melanoma

- The aim of treating this group of patients is only to afford a reasonable palliation. More than 50% of patients, who have metastasis in the regional nodes are dead by the end of one year.

Modes of treatment

1. Radiotherapy

- Patients who have been deemed inoperable due to severe medical co-morbidities. 5-year survival is 70% but local recurrence is unacceptablly high
- Adjuvant radiation to regional lymph node basin for
  - multiple +ve lymph nodes
  - 1 or more metastatic lymph nodes larger than 3 cm
  - Extracapsular extension
  - Recurrent regional disease may benefit from adjuvant radiation.

2. **Systemic chemotherapy**—dacarbazine (DTIC-dmethyl triazenyl imidazole carboxamide) is the standard agent. The response rate is around 20–30%. Cisplatin, Vinblastine, Bleomycin are also used in combination. The addition of immunomodulators such as BCG or Levamisole to chemotherapy have been tried.

- ***Temozolomide*** is an oral analogue of dacarbazine. It achieves significant CNS penetration. It may decrease incidence of cerebral metastasis.

3. **Immunotherapy**

- Interferon alfa-2b (Key Box 11.15)
- Patients with locally advanced, recurrent, nodal, in-transit (Fig. 11.58) or satellite disease should be considered for adjuvant high dose IFN alfa-2b.

Fig. 11.58: Melanoma in-transit deposits

KEY BOX 11.15

INTERFERON-α

- It has stimulatory effect on natural killer cells (NK)
- It has anti-angiogenic activity
- It is toxic—weight loss and myelosuppression can occur
- IL_2—can cause capillary leak syndrome—hypotension and renal failure.

- **Monoclonal antibodies:** These are directed against antigen, expressed on the surface of melanoma cells. These antibodies are similar to IgG3 activate the immune system.
- Cutaneous nodules can be managed by surgery or CO_2 laser excision.
- Intralesional injection of BCG has caused regression of the cutaneous nodules in some patients.

4. **Hormone treatment:** Antioestrogens such as tamoxifen have been tried in the systemic disease with 15–20% response rate.

5. **Isolated limb perfusion:** This is tried when there are extensive in-transit deposits in the limb (Fig.11.59) or recurrent disease in the limb. Melphalan is the drug of choice. A tourniquet is applied first, femoral vein and artery are cannulated, the blood which comes out is passed through a pump and **oxygenator**, into which a high dose of Melphalan or DTIC is given. Input temperature is kept at 41°C. Therapy is aimed at controlling local disease in the limb and to give a better functional limb even in presence of metastasis.

Complications of isolated limb perfusion

- Deep vein thrombosis
- Pulmonary emboli
- Complications of anticoagulants
- Damage to the vessels

**See* Key Box 11.16 for interesting mathematics

KEY BOX 11.16

MATHEMATICS IN MALIGNANT MELANOMA

- 1 mm: Thin melanoma
- 1–4: Intermediate
- 4 and above: Thick melanoma
- > 6 mm diameter of a mole: Suspect melanoma
- 5 cm within primary—satellite nodule
- 8 to 12%: Familial variety
- 20 times more common in whites
- 50 to 60%: Arise from benign naevus
- 70%: Superficial spreading melanoma

Fig. 11.59: Dermatofibroma protuberans upper limb

Summary of skin tumours (Table 11.6)

OTHER MALIGNANT SKIN TUMOURS

Dermatofibrosarcoma protuberans

- This is a **locally malignant tumour** arising from the dermis.
- Common sites are—trunk, flexor region of limbs. It presents as nodular (bosselated) ulcerative lesion of many years duration.
- **Regional lymph node involvement** is uncommon.
- It is less aggressive. Hence, it is curable.
- Treatment is by **local wide excision** followed by primary closure or skin grafting.

Kaposi's angiosarcoma
(Key Boxes 11.17 and 11.18)

- Common in **black population**

KEY BOX 11.17

DISEASES ASSOCIATED WITH KAPOSI'S SARCOMA

- Diabetes mellitus
- Lymphoma
- Following renal transplantation
- Acute and chronic immunosuppression (HIV)

KEY BOX 11.18

TYPES OF KAPOSI'S SARCOMA

- European : Elderly males
- African : Young and children
- Transplant : Due to immunosuppression
- AIDS : Homosexuals

Table 11.6 Summary of skin tumours

	Squamous cell carcinoma	Basal cell carcinoma	Malignant melanoma
Incidence	Common (20–30%)	Most common (60–70%)	Less common (10–20%)
Origin	Prickle cell layer	Basal layer	Melanoblasts
Aetiology	Chronic irritation	Ultraviolet rays, fair skin	Ultraviolet rays, fair skin, pre-existing mole
Site	Trunk, leg, hand, oral cavity	Tear cancer	Head and neck, face, digits, palm and sole
Types	Ulcerative or cauliflower	Nodular or ulcerative	Nodular or ulcerative
Edge	Everted	Rolled out and beaded	Irregular
Induration	Maximum	Moderate	Minimum
Scab	Never occurs	Occurs	Never occurs
Pigmentation	Absent	Absent	Present in 90% of cases
Spread	Mainly by lymphatics. Blood spread is rare and late	Does not spread by lymphatics Blood spread is very, very late Spreads by local spread. Hence the name, rodent ulcer	Mainly by lymphatics, also by blood spread, does not infiltrate like rodent ulcer
Cytokeratin	Expression	No	No

- This neoplasm arises from **proliferating capillary vessels** and perivascular connective tissue cells.
- **Multiple, purplish nodules** appear in the limb, which ulcerate and bleed. This is its characteristic feature.
- Regional lymph node involvement can occur
- Increasing incidence due to AIDS.

Differential diagnosis

1. Malignant melanoma
2. Soft tissue sarcoma
3. Multiple cutaneous metastasis
4. T cell lymphoma

OTHER SKIN LESIONS

They arise from sebaceous glands, sweat glands, hair follicles, etc. *See* Key Box 11.19 for types of exocrine glands.

- Few examples are syringoma, hidradenoma, trichoepithelioma and sebaceous carcinoma.
- They present as localised swelling treated by excision.
- **They have to be kept in mind as a differential diagnosis for malignant skin tumours**. Details of a few skin lesions are given in this chapter.

KEY BOX 11.19

TYPES OF EXOCRINE GLANDS

- ***Holocrine:*** Entire cell dies or disintegrates to liberate secretion, e.g. sebaceous gland.
- ***Apocrine:*** Only the luminal part of the cell disintegrates, cell regeneration takes place from the nucleus and basal portion, e.g. mammary gland.
- ***Merocrine:*** Secretion is discharged without destruction of the cells. Most of the glands belong to this type.

KERATOACANTHOMA: Molluscum sebaceum, Molluscum pseudocarcinomatosum

- Self-limiting benign neoplasm of viral origin (probably)
- Arises due to overgrowth of hair follicle and subsequen spontaneous regression is characteristic.
- It is painless swelling in the skin with central dark brow core. After initial rapid growth of 2–4 weeks, spontaneou regression occurs in 24 hours. After separation of th central core, lump diminishes in size leaving a dee indrawn scar.
- Usually single, face is the commonest site.
- Like sebaceous cyst, it presents as hemispherical swelling
- Treated by excision.
- Keratoacanthoma if associated with sebaceous carcinoma and visceral malignancy (colon cancer) constitutes Muir-Torre syndrome (Fig. 11.62).

TURBAN TUMOUR (Figs 11.60 and 11.61)

It is the **blanket term** used to describe a tumour occupying the whole of the scalp resembling a **turban**.

- Most often used to describe multiple cylindromata
- They produce **pink nodular masses**

KEY BOX 11.20

TURBAN TUMOUR

- Very, very rare

Types

- Multiple cylindromata
- Multiple nodular basal cell carcinoma
- Hidradenomata
- Plexiform neurofibromatosa of scalp

Fig. 11.60: Turban tumour due to squamous cell carcinoma of scalp

Fig. 11.61: Turban tumour (*Courtesy:* Dr Sreejayan, Professor of Surgery, Calicut Medical College)

Fig. 11.62: Sebaceous carcinoma

- Diagnosis is confirmed by biopsy.
- For differential diagnosis *see* Key Box 11.20.
- Treatment includes excision and reconstruction by skin grafting or rotational flaps.

CORN

It is a **popular painful lesion in the plantar surface of the foot (sole of the foot)**.

- It affects the plantar surface of toes and sole of the feet.
- Corn develops due to intermittent pressure over a limited area.
- Basically it is a localised hyperkeratinisation of the skin with a hard central core.
- It is a cone-shaped lesion with broad surface and narrow at deeper plane.
- They are **painful and very tender**.
- Most of these are **hard corns**.
- **Soft corn** can occur in between the toes.

Treatment

- Diabetic patients have to be carefully explained the consequences of a 'mistreated' corn. Sensations and pulsation have to be checked in those patients.
- Symptomatic corns have to be excised. **Excision of a good cone-shaped** tissue is necessary for a permanent cure. Otherwise, **recurrence can occur**.

WART

- A wart is a rough excrescence on the skin
- Papillomaviruses are responsible for this
- They are pigmented, keratinised, irregular lesions
- Common in young adults
- Common sites: Fingers, feet, genitalia, beard area, etc.
- **Veneral warts:** They are also called **papilloma acuminata**. They can occur in the anal region, perineum and in the coronal sulcus of the penis. Some of the warts may regress spontaneously. **Fulguration** with diathermy is the treatment.

MERKEL CELL CARCINOMA

- It is derived from neuroendocrine cells which function as touch receptors.
- **Highly malignant tumour**
- Elderly white males are affected.
- Sun-affected areas such as head and neck regions are involved probably due to **ultraviolet rays**.
- Surgery, radiation and chemotherapy have been tried.
- Histopathological report resembles **metastatic oat cell carcinoma**.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- Treatment of basal cell carcinoma has been discussed in more detail including CO_2 laser excision. Reconstruction with various flaps have been discussed, in more detail.
- A few lines have been added for malignant melanoma including α-interferon.
- A few pearls of wisdom have been added.

MULTIPLE CHOICE QUESTIONS

1. Which of the following statements is false about basal cell carcinoma?
A. It starts as a nonhealing ulcer of many months duration
B. Outer canthus of the eye is the common site
C. It does spread by lymphatics
D. Haematogenous spread is almost unknown

2. Following are true for basal cell carcinoma *except*:
A. It is the most common malignant tumour
B. It cannot occur in the lips
C. Arsenic used in skin ointments may increase the risk
D. Morpheic variety grows very slowly

3. Characteristic feature of basal cell carcinoma is:
A. Epithelial pearls
B. Mitotic figures
C. Orphan Annie nucleai
D. Pallisading islands

4. Following are used to treat basal cell carcinoma *except:*
A. Wide excision
B. Mohs' micrographic surgery
C. Radiation
D. Radiofrequency ablation

5. Following are true for basal cell carcinoma *except*:
A. Ultraviolet rays can predispose to this condition
B. Mohs' micrographic surgery can minimise the recurrence
C. CO_2 laser can be used to treat deep lesions
D. Gorlin's syndrome is associated with multiple basal cell carcinoma

6. Following skin malignancies can spread by lymphatic spread *except:*
A. Epithelioma
B. Malignant melanoma
C. Sebaceous carcinoma
D. Basal cell carcinoma

7. Characteristic feature of epithelioma is:
A. Penetrating lesion
B. Elevated beaded edge
C. Proliferative growth with everted edges
D. Nodular lesion

8. Following are true for epithelioma *except:*
A. Majority are well differentiated
B. Cell nest is characteristic
C. Broder's classification is used
D. Polyhedral cells

9. Following are true for Marjolin's ulcer *except:*
A. It arises from scar tissues
B. It does not spread by lymphatics
C. It is rapidly growing
D. It is painless

10. Following are true about prognostic factors fo epithelioma *except*:
A. Metastasis is unlikely if the lesion is more tha 10 mm
B. Broder's high grade means poor prognosis
C. Recurrence is more in lip
D. Perineural involvement carries poor prognosis

11. About melanocytes which one of the following is false
A. They are derived from neural crest
B. They convert DOPA into melanin
C. Melanocyte stimulating hormone is released fron intermediate lobe of pituitary gland
D. Present in the stratum basale layer of the epidermis

12. Following are true for malignant melanoma *except:*
A. Spindle cell naevus has high malignant potential
B. Junctional naevus has very high malignant potential
C. Superficial spreading variety is the most common form
D. Tyrosinase is absent in albinos

13. Following are true for thin melanomas *except:*
A. Less than 1 mm thickness
B. Metastasis is about 10–15 %
C. 20 year survival is almost 95%
D. Local wide excision of 2 cm margin is recommended

14. Following are true for acral lentigenous *except:*
A. Least common subtype
B. Occur in palm and soles
C. Hutchinson's sign may be found
D. Rarely found in Blacks

15. Following are true for superficial spreading type malignant melanoma *except:*
A. Most common subtype
B. Trunk is the common site
C. Average age of presentation is 5th decade
D. It has a long vertical growth

16. Which one of the following treatment is recommended in 2 cm melanoma leg with inguinal nodes?
A. Wide excision with inguinal block dissection
B. Wide excision with ilioinguinal block dissection
C. Wide excision with ilioinguinal obturator dissection
D. Amputation and inguinal block dissection

17. Which one of the following drugs used to decrease cerebral metastasis in malignant melanoma?
A. Adriamycin
B. Cisplatin
C. Temozolamide
D. Alpha-interferon

18. Alpha-interferon has following actions *except:*
A. It has stimulatory effect on natural killer cells
B. It has anti-angiogenic activity
C. Does not cause myelosuppression
D. Can cause capillary leak syndrome

19. Following are true for Merkel cell tumour *except:*
A. It is a highly malignant tumour
B. Derived from neuroendocrine tumours
C. Fingers and digits are the common sites
D. Head and neck are commonly involved

20. Following are true for Kaposi's angiosarcoma *except:*
A. Common in Black population
B. Paclitaxel is the drug of choice with a 75% effective rate
C. Homosexuals are affected more as a result of AIDS
D. Classic form affects head and neck more than legs

ANSWERS

1 C	2 D	3 D	4 D	5 C	6 D	7 C	8 D	9 C	10 A
11 C	12 B	13 D	14 D	15 D	16 B	17 C	18 C	19 C	20 D

12

Haemorrhage, Shock and Blood Transfusion

- Haemorrhage
  - Classification
  - Pathophysiology
  - Management
- Shock
  - Classification
  - Pathophysiology
  - Management
- Acute adrenal insufficiency
- Hyperbaric oxygen
- Central venous pressure
- Pulmonary capillary wedge pressure
- Blood transfusion
- Blood products
- Plasma substitutes
- Bleeding disorders

HAEMORRHAGE

Classification

I. Depending on nature of the vessel involved

A. Arterial haemorrhage: Bright red in colour, jets out. Pulsation of the artery can be seen. It can be easily controlled, as it is visible.

B. Venous haemorrhage: Dark red in colour. It never jets out but oozes out. It is nonpulsatile and difficult to control because vein gets retracted.

C. Capillary haemorrhage: Red in colour, never jets out, slowly oozes out. It becomes significant if there are bleeding tendencies.

II. Depending on the timing of haemorrhage

A. Primary haemorrhage: Occurs at the time of surgery

B. Reactionary haemorrhage: Occurs after 6–12 hours of surgery. Hypertension in the postoperative period, violent sneezing, coughing or retching, are the usual causes, e.g. superior thyroid artery can bleed after thyroidectomy, if the ligature slips. Hence, it is better to ***ligate it twice***.

C. Secondary haemorrhage: Occurs after 5–7 days of surgery. It is due to infection which eats away the suture material, causing sloughing of vessel wall, e.g. bleeding after 5–7 days of surgery for haemorrhoids.

III. Depending on the duration of haemorrhage

A. Acute haemorrhage: Occurs suddenly, e.g. oesophageal variceal bleeding due to portal hypertension.

B. Chronic haemorrhage: Occurs over a period of time, e.g. haemorrhoids/piles or chronic duodenal ulcer, tuberculous ulcer of the ileum, diverticular disease of the colon.

IV. Depending on the nature of bleeding

A. External haemorrhage/revealed haemorrhage, e.g. epistaxis, haematemesis.

B. Internal haemorrhage/concealed haemorrhage, e.g. splenic rupture following injury, ruptured ectopic gestation, liver laceration following injury.

Pathophysiology of haemorrhagic shock

A loss of more than 30–40% blood volume results in a fall in blood pressure and gross hypoperfusion of the tissues leading

CLINICAL NOTES

A patient who underwent subtotal thyroidectomy for toxic goitre was shifted to the postoperative intensive care unit. Within 10 minutes, the nurse came to inform the surgeon that 450 ml of blood was collected in the "Redivac" suction bottle. The dressing was opened and there was no large collection of blood in the surgical wound. The presence of a large haematoma requiring drainage was thus ruled out. The blood pressure (BP) which was previously under control had shot up to 210/110 mmHg postoperatively, possibly due to pain. Careful monitoring and treatment reduced the BP to 140/90 mmHg. After 24 hours, the drainage was only 100 ml. The incision did not need re-exploration. This case illustrates reactionary haemorrhage due to hypertension.

to haemorrhagic shock. **The evolution of haemorrhagic shock can be classified into four stages.**

Class I

- When blood loss is less than 750 ml (< 15% of blood volume), it can be called mild haemorrhage.
- 60–70% of blood volume is present in the low-pressure venous system (capacitance vessels). Ten per cent of the blood volume is present in the splanchnic circulation.
- When there is blood loss, peripheral venoconstriction takes place and compensates for the loss of blood volume by shifting some blood into the central circulation. Some amount of correction of blood volume also occurs due to withdrawal of fluid from the interstitial spaces.
- Apart from a mild tachycardia and thirst, there may be no other symptom or sign suggesting hypovolaemia. The blood pressure, urine output and mentation are all normal in Class I shock.

Class II

- Loss of 800–1500 ml (15–30% of blood volume) results in moderate (Class II) shock.
- Peripheral venoconstriction may not be sufficient to maintain the circulation. Hence, adrenaline and noradrenaline (catecholamines) released from the sympathoadrenal system cause powerful vasoconstriction of both arteries and veins.
- Increased secretion of ADH causes retention of water and salt. Thirst increases.
- Clinically, the patient shows a heart rate of 100–120 beats/minute and an elevated diastolic pressure. The systolic pressure may remain normal. Urine output is reduced to about 0.5 ml/kg/h and the capillary refill is more than the normal 2 seconds. Extremities may look pale and the patient is confused and thirsty.

Class III

- Loss of 1500–2000 ml (30–40% of blood volume) produces Class III shock. All the signs and symptoms of Class II haemorrhage get worse.
- The patient's systolic and diastolic blood pressures fall and the heart rate increases to around 120 beats/minute. The pulse is thready.
- The respiratory rate increases to more than 20/minute. Urine output drops to 10 to 20 ml/hour. The patient appears pale and is aggressive, drowsy or confused.

Class IV

- A blood loss of more than 2000 ml (> 40% of blood volume) results in Class IV shock. The peripheries are cold and ashen.
- The pulse is thready and more than 120/minute. The blood pressures are very low or unrecordable.
- The patient may be moribund.
- If persistent, can damage other organs, e.g.

  GIT: Mucosal ulcerations, upper GI bleeding, absorption of bacteria and toxins, **bacterial translocation** and bacteraemia

  Liver: Reduced clearance of toxins

  Kidney: **Acute renal failure**

  Heart: **Myocardial ischaemia**, depression

Lungs: Loss of surfactant, increased alveolocapillary permeability, interstitial oedema, increased arteriovenous shunting results in **acute lung injury** (ALI).

Multiorgan failure consequent to haemorrhagic shock is associated with a high mortality rate.

Early diagnosis of bleeding and appropriate management is crucial to improve survival and outcome.

Management of haemorrhagic shock

I. Treatment—general measures

- Hospitalisation
- Care of all critically ill patients begins with **A**, **B** and **C**. A: Airway, B: Breathing, and C: Circulation.
- **Oxygen** should be administered by face mask to all patients who are in shock but are conscious and are able to maintain their airway.
- If unconscious, endotracheal intubation and ventilation with oxygen may be necessary.
- Haemorrhage control
- **Intravenous access:** Urgent intravenous administration of Ringer lactate to restore blood volume to normal. If there has been massive blood loss as in Class IV shock or the patient is anaemic, blood transfusion is indicated. Colloids such as gelofusine or 5% albumin may also be used. The use of hetastarch may be associated with increased rate of acute kidney injury and mortality, and hence better avoided.
- **Investigations:** Blood is collected at the earliest opportunity for routine investigations as well as for blood grouping and cross-matching.
- **Cross-matched blood is usually given:** If the haemorrhage is life-threatening, uncross-matched, O –ve packed cells may be transfused into the patient.
- Use of inotropes and vasoconstrictors is not indicated as they may harm tissue perfusion.
- However, if inotropes have been started as a life-saving measure, an attempt should be made to wean them as soon as the volume status is corrected and the patient is stable.

II. Treatment—specific measures

1. Pressure and packing

- To control bleeding from nose and scalp: Packing using roller gauze with or without adrenaline to control bleeding from nose.

- Bleeding from vein: Middle thyroid vein during thyroidectomy, lumbar veins during lumbar sympathectomy can be controlled using pressure pack for a few minutes.
- Sengstaken tube is used to control bleeding from oesophageal varices—internal tamponade.

2. Position and rest

- Elevation of the leg controls bleeding from varicose veins
- Elevation of the head-end reduces venous bleeding in thyroidectomy—**anti-Trendelenburg position**.
- Sedation to relieve anxiety—Midazolam in titrated doses of 1–2 mg intravenously may be given.

3. Tourniquets

Indications

- Reduction of fractures
- Repair of tendons
- Repair of nerves
- When a bloodless field is desired during surgery

Contraindications

Patient with peripheral vascular disease. (The arterial disease may be aggravated due to thrombosis resulting in gangrene.)

Types

- Pneumatic cuffs with pressure gauge
- Rubber bandage

Precautions

- ***Too loose*** a tourniquet does not serve the purpose.
- ***Too tight***: Arterial thrombosis can occur which may result in gangrene.
- ***Too long*** (duration of application): Gangrene of the limb. Hence, when a tourniquet is applied, the time of inflation should be noted down and at the end of 45 minutes to an hour, it has to be deflated at least for 10 minutes and reinflated only if necessary.

Complications

- Ischaemia and gangrene
- Tourniquet nerve palsy[1]

4. Surgical methods to control haemorrhage

- Application of **artery forceps** (Spencer Well's forceps) to control bleeding from veins, arteries and capillaries.
- Application of **ligatures** for bleeding vessels.
- **Cauterisation** (diathermy).
- Application of **bone wax** (**Horsley's wax** which is bee's wax in almond oil) to control bleeding from cut edges of bones.
- **Silver clips** are used to control bleeding from cerebral vessels ***(Cushing's clip)***.
- Surgical procedure: **Splenectomy** for splenic rupture hysterectomy for uncontrollable postpartum haemorrhage laparotomy for control of bleeding from ruptured ectopi pregnancy.

SHOCK

Definition

Shock is defined as an acute clinical syndrome characterise by **hypoperfusion and severe dysfunction of vital organs** There is a failure of the circulatory system to supply blood i **sufficient quantities** or under **sufficient pressure** necessar for the optimal function of organs vital to survival.

Classification

- Hypovolaemic shock
- Cardiogenic shock
- Distributive shock
- Obstructive shock

HYPOVOLAEMIC SHOCK

- Loss of blood—haemorrhagic shock
- Loss of plasma—as in burns shock
- Loss of fluid—dehydration as in gastroenteritis

Features (Key Box 12.1)

The primary problem is a decrease in preload. The decreased preload causes a decrease in stroke volume.

Clinical features depend on the degree of hypovolaemia. Severe (Class III or IV) shock results in tachycardia, low blood pressures and decreased urine output.

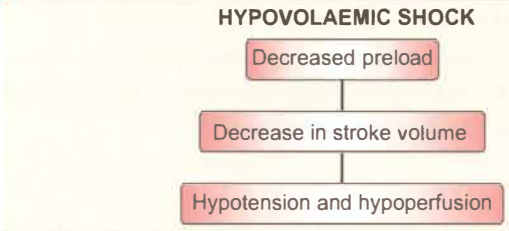
The peripheries are cold and the patient may be confused or moribund (*see* pathophysiology of haemorrhagic shock).

Treatment

- The primary goal is to restore the blood volume, tissue perfusion and oxygenation to normal as early as possible.
- Replace the lost blood volume.

KEY BOX 12.1

HYPOVOLAEMIC SHOCK



[1]In MS examination, a candidate was asked to examine a case of radial nerve palsy. The patient had an injury to the wrist 4 months back. The cut flexor tendons had been sutured. The candidate could not correlate the radial nerve palsy to the injury at the wrist. He failed! It was a case of tourniquet palsy.

- ***Crystalloids:*** If crystalloids are used to replace blood loss, 2–3 times the volume lost needs to be given. Ringer lactate is the crystalloid of choice. Large volumes of saline infusion can cause hyperchloraemic metabolic acidosis. 5% dextrose is not used to expand the intravascular volume as it is hypotonic once the dextrose is metabolised.
- ***Colloids:*** When colloids are used to replace lost blood volume, a volume equal to the lost volume may be given.
- Crystalloids are preferred in the initial phase of resuscitation. If large volume of blood is lost, colloids such as gelofusine or 5% albumin may be given in order to reduce the amount of volume to be infused.
- ***Blood transfusion*** may be needed if large amount of blood is lost (> 40% of blood volume) or if the patient is anaemic (Hb < 8 g%).

CARDIOGENIC SHOCK

The blood flow is reduced because of an intrinsic problem in the heart muscle or its valves. A massive myocardial infarction may damage the cardiac muscle so that there is not much healthy muscle to pump blood effectively. Any damage to the valves, especially acute may also reduce the forward cardiac output resulting in cardiogenic shock.

Features

- The primary problem is a decrease in contractility of the heart. The decreased contractility causes a decrease in stroke volume.
- Left ventricular pressures rise as forward cardiac output reduces. The sympathetic nervous system is activated and consequently, systemic vascular resistance increases.
- Clinically, the patient presents with tachycardia, low blood pressures and decreased urine output.
- The jugular venous pulse may be raised, a S_3 or S_4 gallop may be present.
- The lung fields may show bilateral extensive crepitations due to pulmonary oedema.
- The peripheries are cold and the patient may be confused or moribund.

Treatment

- The primary goal is to **improve cardiac muscle function**.
- Oxygenation can be improved by administering oxygen, either by face mask or by endotracheal intubation and ventilation as necessary.
- **Inotropes** improve cardiac muscle contractility.
- **Vasodilators** such as nitroglycerine may dilate the coronary arteries and peripheral vessels to improve tissue perfusion. Lowering systemic vascular resistance reduces impedance to forward cardiac output (afterload). However, the patient must be monitored closely to avoid excessive drops in blood pressure due to these drugs.
- **Intra-aortic balloon pump** or ventricular assist devices may be used to augment cardiac output.
- If hypotension continues to be unresponsive, revascularisation (surgical or interventional) or valve replacements may be considered on an emergency basis.

DISTRIBUTIVE SHOCK

In distributive shock, the afterload is excessively reduced affecting circulation. Distributive shock can occur in the following situations:

- Septic shock
- Anaphylactic shock
- Neurogenic shock
- Acute adrenal insufficiency

SEPTIC SHOCK

Pathophysiology

- Sepsis is the response of the host to bacteraemia/endotoxaemia.
- It may be produced by gram-negative or gram-positive bacteria, viruses, fungi or even protozoal infections.
- Severe sepsis can result in persistent hypotension despite adequate fluid resuscitation and is called septic shock.
- Local inflammation and substances elaborated from organisms, especially endotoxin, activate neutrophils, monocytes and tissue macrophages. **This results in a cascade of proinflammatory and anti-inflammatory cytokines and other mediators such as IL-1, IL-8, IL-10, tumour necrosis factor-alpha, prostaglandin E_1, endogenous corticosteroids and catecholamines.**
- Effects of this complex mediator cascade include cellular chemotaxis, endothelial injury and activation of the coagulation cascade (Key Box 12.2).

Features

- These substances produce **low systemic vascular resistance (peripheral vasodilatation)** and ventricular dysfunction resulting in persistent hypotension.
- **Generalised tissue hypoperfusion** may persist despite adequate fluid resuscitation and improvement in cardiac

KEY BOX 12.2

DISTURBANCE OF PROCOAGULANT—ANTICOAGULANT BALANCE

- Inflammatory response in sepsis activates tissue factor which in turn activates coagulation
- Fibrinogen is converted to fibrin
- Lowered levels of natural anticoagulants such as protein C, protein S and antithrombin III
- Procoagulant—anticoagulant imbalance → diffuse microvascular thrombi

output and blood pressures. This is due to abnormalities in regional and microcirculatory blood flow. These abnormalities may lead to **cellular dysfunction, lactic acidosis (anaerobic metabolism) and ultimately, multi-organ failure**.

- Early phases of septic shock may produce evidence of volume depletion such as dry mucous membranes and cool, clammy skin.
- After resuscitation with fluids, however, the clinical picture is typically more consistent with **hyperdynamic shock.** This includes tachycardia, bounding pulses with a widened pulse pressure, a hyperdynamic precordium on palpation and warm extremities.
- Signs of possible infection include fever, localised erythema or tenderness, consolidation on chest examination, abdominal tenderness, guarding, rigidity and meningismus.
- Signs of **end-organ hypoperfusion** include tachypnoea, cyanosis, mottling of the skin, digital ischaemia, oliguria, abdominal tenderness and altered mental status.
- Often, a definitive diagnosis cannot be made on the basis of initial findings, on history taking and physical examination and empirical treatment for several possible conditions commences simultaneously.

Treatment

- **Removal of the septic focus is an essential step** and a priority in the treatment of septic shock, e.g. resection of gangrenous bowels, closure of perforation, appendicectomy.
- Early (preferably given in the first hour of arrival) empirical **antibiotic therapy** to treat the precipitating infection.
- **Supportive care:** Oxygenation and if necessary, endotracheal intubation and mechanical ventilation should be administered.
- **Intravenous fluids:** Restoration of intravascular filling pressures must be done using crystalloids, colloids and blood as necessary. **Crystalloids such as isotonic saline** or **Ringer's lactate** may be used. Large amounts may be required and may contribute to tissue oedema. Colloids restore intravascular volume faster and remain longer in the central circulation. However, they are expensive and are more often used in patients where there is a high-risk of pulmonary oedema due to cardiac dysfunction and thus, may not tolerate large volume of fluids. Avoid hetastarch as it may increase risk of renal injury and mortality. **Blood transfusions** may be required to maintain the patient's haemoglobin levels at 8–10 g%.
- **Vasoactive agents** such as norepinephrine to produce vasoconstriction and raise the systemic vascular resistance to normal may be used. Dopamine, dobutamine or adrenaline may need to be added. Vasopressin infusion may be useful in patients with refractory shock. All these potent drugs are given as infusions under careful and continuous monitoring of blood pressure as well as cardiac filling pressures (central venous pressures).

PEARLS OF WISDOM

No matter what antibiotics you use, unless you remove the septic focus by surgical drainage of pus or resection of gangrene, etc. patients in septic shock do not improve.

Summary of septic shock (Key Box 12.3)

KEY BOX 12.3

SEPTIC SHOCK

- Early diagnosis of septic shock
- Empirical antibiotics initially
- Appropriate antibiotics after culture and sensitivity testing
- Ultrasonography, CT scan, and chest X-ray are key investigations
- **Treatment of source of infection**
  - Pneumonia
  - Drainage of pus
  - Closure of perforation
  - Resection of gangrene
- Early and aggressive resuscitation, supportive care and close monitoring in intensive care unit (ICU).

ANAPHYLACTIC SHOCK

Features

Occurs on exposure to an allergen the patient is sensitive to. It may be pollen, foodstuffs, preservatives in the food or a medication. The anaphylactic shock that occurs in the hospital is usually due to some drug, e.g. the patient is allergic to penicillin. Latex allergy is being increasingly recognised.

The reaction may be in the form of **mild rashes** with or without **bronchospasm** or it may be a full blown anaphylactic shock wherein the patient presents with rashes, **generalised oedema** including laryngeal oedema, **bronchospasm** and **hypotension** and if not treated in time, cardiac arrest.

Treatment

I. Primary

- Oxygen and if necessary, endotracheal intubation and ventilation.
- Adrenaline, 0.5–1 mg IM or 50–100 µg intravenous boluses as necessary to maintain blood pressure.
- Intravenous fluids—isotonic saline or Ringer lactate
- Leg end elevation of bed.

II. Secondary

- Chlorpheniramine maleate
- Hydrocortisone 100 mg intravenously
- If facilities exist, take a 10 ml sample of blood to analyse for serum tryptase levels. If raised, they confirm anaphylactic reaction.

NEUROGENIC SHOCK

Causes: High spinal cord injury, vasovagal shock

Features: Hypotension without tachycardia that can deteriorate to produce shock and cardiac arrest.

Treatment: Intravenous fluids, inotropes and vagolytics as necessary.

ACUTE ADRENAL INSUFFICIENCY

Causes (Key Box 12.4)

Adrenal crisis occurs if the adrenal gland is deteriorating as in:

- Primary adrenal insufficiency (Addison's disease)
- Secondary adrenal insufficiency (pituitary gland injury, compression)
- Inadequately treated adrenal insufficiency.

Features

- Headache, profound weakness, fatigue, slow and lethargic movement, joint pain.

KEY BOX 12.4

RISK FACTORS FOR ADRENAL CRISIS

- Infection
- Trauma or surgery
- Adrenal gland or pituitary gland injury
- Premature termination of treatment with steroids such as prednisolone or hydrocortisone.

- Nausea, vomiting, abdominal pain, high fever and chills
- Low blood pressure, dehydration, rapid heart rate and respiratory rate, confusion or coma.

Treatment

- Care of airway, breathing and circulation
- Intravenous fluids
- Hydrocortisone 100–300 mg intravenously
- Treat the precipitating factor
- Antibiotics as necessary

OBSTRUCTIVE SHOCK

It can be due to cardiac tamponade or due to tension pneumothorax.

A. CARDIAC TAMPONADE

Features

- In obstructive shock, there is impedance to either inflow or outflow of blood into or out of the heart.

CLINICAL NOTES

A 54-year-old lady was admitted to the casualty with low blood pressures and dyspnoea since one day. She had a history of fever, vomiting and diarrhoea since 3–4 days, was treated in a local nursing home and when she got worse, was referred to our hospital. In spite of fluid therapy, profound hypotension persisted and within half an hour of arrival to the casualty, she suffered a cardiopulmonary arrest.

Her trachea was immediately intubated, cardio-pulmonary resuscitation (CPR) given and was shifted to the intensive care unit for further management. She required high doses of dopamine, adrenaline and noradrenaline to maintain blood pressures. A blood gas analysis showed severe metabolic acidosis (pH = 7.02, $PaCO_2$ = 35 mmHg and HCO_3^- = 12 mmol/L).

Considering the history, a diagnosis of septic shock was made when she continued to have hypotension even after her central venous pressures were normal. Peritoneal dialysis was done as she was in oliguric renal failure. Haemodialysis was not possible as she was hypotensive and on inotropes. A search for septic focus was initiated. An ultrasound abdomen showed dilated kidney and obstructed urinary system.

A double J stenting of the ureter, which was done to relieve the obstruction, drained pus. Once the pus was drained, appropriate antibiotics were given and with continued cardiorespiratory support, she showed steady improvement. She was gradually weaned off the ventilator and inotropes, and was discharged from the hospital five weeks later. At discharge, she was fully conscious, stable, ambulant and very grateful to the medical fraternity.

This case illustrates the importance of resuscitation, cardiorespiratory support, removal of septic focus and antibiotics in the treatment of septic shock.

- In **cardiac tamponade,** the pericardium is filled with blood and hampers venous filling as well as outflow.
- The filling pressures of the left-sided and right-sided chambers equalise.
- **The central venous pressure is high and the blood pressure is low.**
- The patients also have pulsus paradoxus where there is 10% decrease in systolic blood pressure with inspiration.

Treatment

- To maintain preload with fluids or blood as indicated.
- Relief of obstruction, drain the pericardial cavity as early as possible.

B. TENSION PNEUMOTHORAX

Causes

- Injury to the lung due to trauma
- Ventilator-induced barotrauma
- Rupture of emphysematous bulla in a patient with chronic obstructive pulmonary disease.

Features

- Profound cyanosis, distended neck veins
- Tachypnoea, dyspnoea or respiratory arrest
- No air entry on the side of pneumothorax, hyper-resonance to percussion
- Tachycardia, hypotension and cardiac arrest

Treatment

- A wide (large) bore needle/cannula must be inserted into the pleural cavity to drain the air. The needle is inserted in the midclavicular line in the 2nd intercostal space on the affected side. This is followed by insertion of a tube thoracostomy.

PEARLS OF WISDOM

A massive pulmonary embolus is a differential diagnosis for obstructive shock.

HYPERBARIC OXYGEN

Hyperbaric oxygen is administered to patients when their own haemoglobin is unable to carry adequate oxygen or when hyperoxia is required. Hyperbaric O_2 is administered using either a monoplace (single patient) or multiplace compression chamber.

Indications

1. **Carbon monoxide poisoning:** Hyperbaric O_2 at 2–3 atm is given to increase the O_2 dissolved in plasma (5–6 ml/dl at 3 atm, whereas it is 0.3 ml/dl at 1 atm) which is sufficient to meet the O_2 requirement of the body even in the absence of normal haemoglobin (severe CO poisoning).
2. Infections such as **gas gangrene**: Hyperoxia suppresses the growth of anaerobic organisms.
3. Cancer therapy to potentiate radiotherapy
4. Arterial insufficiency
5. **Decompression sickness and air embolism:** Hyperbaric oxygen reduces the size of air bubbles and rapidly eliminates nitrogen from the air bubbles.

Contraindications

Untreated pneumothorax: Can expand with hyperbaric O_2 resulting in life-threatening tension pneumothorax.

CENTRAL VENOUS PRESSURE (CVP)

- One of the desirable requirements while treating patients in shock includes monitoring of CVP.
- CVP is a fair indicator of blood volume. Thus, in shock, measurement of CVP is useful so as to plan proper fluid management.
- CVP is also affected by contractility of the right ventricle, intrathoracic pressure and intrapericardial pressure.

Method

- Internal jugular vein (IJV) or subclavian vein are preferred to gain access to central veins (Key Box 12.5).
- A 16 cm long IV catheter is introduced into the central vein with the patient supine, head down and neck rotated to the opposite side. Head down position helps in engorging the vein. Seldinger's technique is employed and the catheter is advanced up to the junction of superior vena cava and right atrium.
- The patency and position of the catheter is confirmed by lowering the saline bottle to check free flow of blood into the connecting tube.
- The tube is connected to a saline manometer. Readings of the saline level are taken with the 'zero reference point' at the **midaxillary level** if the patient is in **supine position** or at the **manubriosternal joint,** if he is in the semireclining position (45°). An electronic pressure transducer may be used for greater accuracy.
- CVP must be measured at end-expiration as it may change with phases of respiration.

Uses

1. If CVP is low, venous return should be supplemented by IV infusion, as in cases of hypovolaemic shock.
2. When CVP is high, further infusion of fluids may result in pulmonary oedema.
3. In cardiogenic shock, CVP may be normal or high.

Complications

1. Pneumothorax
2. Accidental carotid artery puncture
3. Haematoma in the neck
4. Bleeding
5. Air embolism
6. Infection.

KEY BOX 12.5

ACCESS TO RIGHT HEART/GREAT VEINS

- Internal jugular vein
- Subclavian vein
- Median cubital vein
- External jugular vein

PULMONARY CAPILLARY WEDGE PRESSURE (PCWP)

- This is a better indicator of circulatory blood volume and left ventricular function.
- It is measured by a pulmonary artery balloon floatation catheter—**Swan-Ganz** catheter.

Uses of PCWP

1. To differentiate between left and right ventricular failure
2. Pulmonary hypertension
3. Septic shock
4. Accurate administration of fluids, inotropic agents and vasodilators.

Method of measuring PCWP

Swan-Ganz catheter is introduced into the right atrium through a central vein (RIJV usually). The catheter has a balloon near its tip which is inflated with air. The catheter is advanced and pressure tracing is monitored. Its entry into the right ventricle and pulmonary artery is identified by changes in pressure tracing. With further advancement, the pressure tracing becomes flat, when the balloon gets wedged in a small branch to give pulmonary capillary wedge pressure (PCWP). When the balloon is deflated, the pulmonary artery pressure is obtained. **Normal values: PCWP: 8–12 mmHg.** PAP: systolic 25 mmHg; PAP diastolic 10 mmHg.

Complications

- Arrhythmias
- Pulmonary infarction
- Pulmonary artery rupture

Interpretation in various conditions: CVP and PCWP (Table 12.1)

Note: **CVP reflects right atrial pressures only.** It is low in hypovolaemia and, high in hypervolaemia and in right ventricular failure.

PCWP is a better indicator of left ventricular pressures and is a preferred monitor in cardiogenic shock. PCWP is low in hypovolaemia and high in hypervolaemia as well as in left ventricular failure.

Table 12.1

Conditions	CVP	PCWP
Hypovolaemic shock	Low	Low
Right heart failure	High	Normal
Left heart failure	Normal	High
Cardiac tamponade	High	High
Pulmonary embolism	Normal/ High	High

Note: The use of pulmonary artery catheter has declined significantly due to increased complications and unfavourable risk-benefit ratio. An echocardiogram is noninvasive, provides a lot more information and is preferred in the treatment of cardiogenic shock.

BLOOD TRANSFUSION

Administration of whole blood or its components into a patient is often necessary for various reasons. Since the only available source of blood is by voluntary human donation, and that source is scarce, whole blood is separated into its components, namely packed red blood cells, fresh frozen plasma, platelets and cryoprecipitate. Some coagulation factors may be isolated and separately stored, e.g. Factor VIII for administration into haemophiliacs. Packed red cells are the commonest blood products used. Their transfusion increases the oxygen carrying capacity of blood. Transfusion of 250 ml of packed cells raises haemoglobin by 1 g% in an adult.

Indications of blood transfusion (Key Box 12.6)

I. Packed red cells are used to replace acute and major blood loss as in:
   - Haemorrhagic shock
   - Major surgery—open heart surgery, gastrectomy
   - Extensive burns

II. Packed red cells are also used to treat anaemia due to:
   - Extensive burns
   - Chronic blood loss as in haemorrhoids, bleeding disorders, chronic duodenal ulcer, etc.
   - Inadequate production as in malignancies, nutritional anaemia

III. To replace platelets in thrombocytopaenia, fresh frozen plasma in vitamin K deficiency unresponsive to vitamin K replacement as in liver disease or to reverse effects of warfarin, cryoprecipitate to replace fibrinogen in patients with disseminated intravascular coagulation.

IV. Whole fresh blood administration is described in massive trauma with heavy blood loss, the rationale being that the patient rapidly loses all components of blood and so all components need to be replaced. Thus, it makes sense to administer whole blood rather than individual components. However, many blood banks are reluctant to allow whole blood transfusion.

KEY BOX 12.6

GUIDELINES: WHEN TO TRANSFUSE

1. Blood loss > 20% of blood volume
2. Haemoglobin < 8 g/dl
3. Haemoglobin < 10 g/dl in patients with major cardiovascular disease (e.g. ischaemic heart disease)

BLOOD PRODUCTS

Packed red blood cells (PRBC): When whole blood is centrifuged, red blood cells settle down and the platelet rich plasma remains supernatant. The plasma is transferred to another bag while preservative is added to red cells and the bag is sealed. Each bag of packed cells contains approximately 250–300 ml with a red cell concentration of 70%. Red cell transfusion is required for patients whose haemoglobin is < 7 g%, as in patients undergoing chemotherapy, major surgery, delivery, trauma, grossly anaemic neonates and infants or in patients with sickle cell anaemia, especially in sickle cell crisis. A higher transfusion threshold (9–10 g%) may be used for patients with cardiac disease. Each unit of packed cells should increase haemoglobin by 1 g% and haematocrit by 3%.

The red cells may be leucodepleted (white blood cells removed) for use in patients requiring multiple transfusions to prevent development of antibodies to leukocytes or in those who are known to react to leukocytes in the past.

The rate of blood or blood product administration depends on the quantity and speed at which it is lost from the body. Slow transfusion is indicated in patients with cardiac disease, renal dysfunction, severe chronic anaemia and in paediatric patients.

In semi-emergent situations, e.g. treatment of low haemoglobin in patients awaiting surgery or delivery, each bag of packed cells may be transfused slowly. The initial 25 ml can be given slowly to check whether the patient tolerates it. Thereafter, it can be transfused at a rate of 3–4 ml/kg/h (approximately 1.5 h for a bag of red cells). The red cells must be transfused within four hours of removing from refrigerator as bacterial growth may be promoted after that.

In emergent situations, the red cells may be transfused much faster in order to keep up with the loss. When large amounts are transfused in a short period (*see* section on massive transfusion), effects of other components such as citrate become significant.

Packed red blood cells must be both ABO and Rh compatible unless the patient has life-threatening massive bleeding, in which case O –ve packed cells may be transfused.

Platelets: The bag containing platelet rich plasma is again centrifuged to express off the plasma so that the bag with the remaining platelets can be sealed off. Each unit (50 ml) should contain at least 5.5×10^{10} platelets (platelet concentrate) and each unit should elevate the platelet count by 5–10,000 cells/cu mm in a 70 kg person. Platelets may be transfused prophylactically or for therapeutic purposes.

Prophylactic platelet transfusion may be done when platelet count is < 10,000 cells/cu mm in oncology patients. In patients who are at high-risk of alloimmunisation, e.g. leukaemia, the threshold for platelet transfusion is even lower at 5000 cells/cu mm, whereas in patients with clinical instability, the threshold may be raised to 20,000 cells/cu mm. Patients who need to undergo major surgery or requiring invasive procedures such as spinal anaesthesia, liver biopsy, etc. generally need their platelet count elevated to > 50,000 cells/cu mm. However patients requiring surgery in critical areas such as neurosurgery or ophthalmic surgery will need their platelet count raised to > 1,00, 000 cells/cu mm.

Therapeutic platelet transfusion is required in patients who are known to be thrombocytopaenic and are actively bleeding (platelet count < 50,000 cells/cu mm). The dose of platelets may be calculated as follows: One unit of platelets (60 ml) for every 10 kg body weight.

Platelets must be transfused within four hours of commencement of the infusion. ABO compatibility is not required for platelets. Filters of 170–260 μ must be used to transfuse platelets as with other blood components.

Fresh frozen plasma (FFP): The remaining plasma (200–230 ml) may be stored in a frozen form (called fresh frozen plasma) at –18°C for one year. It needs to be thawed over half hour before use. Fresh frozen plasma transfusion is indicated in patients with prolonged INR > 1.5 and are bleeding or require surgery. FFP provides coagulation factors to those patients who are actively bleeding and those on warfarin. Fresh frozen plasma is administered in a dose of 10–20 ml/kg body weight. FFPs must be ABO compatible. FFP must be transfused as soon as possible after thawing and definitely within 24 hours of thawing. A hanging unit must be transfused within six hours of commencement of transfusion due to risk of bacterial contamination.

Cryoprecipitate: The fresh frozen plasma may be further treated to produce cryoprecipitate rich in fibrinogen. Transfusion of cryoprecipitate (each bag contains 20 ml) is indicated in patients whose fibrinogen levels are < 1 g%, e.g. disseminated intravascular coagulation (DIC). The dose is 0.2 units/kg body weight but usually 10 units are transfused initially and repeated as necessary.

Important points to remember before transfusion of blood products:

- Always obtain informed consent from patients or immediate relatives (in emergent situations) prior to transfusion.
- Recheck the patient's blood group and of the donor blood, preferably along with one other qualified person to ensure that the correct bag is being transfused to the patient. The patient's hospital number and the blood bag number should also be checked. Both people must sign on the transfusion report.
- Visually check the blood bag for any obvious abnormalities such as very dark blood, visible clots and if present, do not transfuse but return to blood bank. Check the date of collection and date of expiry before transfusion. The storage shelf life of red blood cells depends on the preservative added. RBCs stored in ACD (acid-citrate-dextrose), CPD (citrate-phosphate-dextrose), CPDA (citrate-phosphate-dextrose-adenine) and SAGM (saline-adenine-glucose-mannitol) solutions have a shelf-life of 21, 28, 35 and 35 days respectively (Key Box 12.7).

KEY BOX 12.7

STORAGE OF BLOOD

Preservative	RBC survival (days)
ACD (acid-citrate-dextrose)	21
CPD (citrate-phosphate-dextrose)	28
CPDA (citrate-phosphate-dextrose-adenine)	35
SAGM (saline-adenine-glucose-mannitol)	35

- Watch for any transfusion reactions.

Whole blood transfusion is not advisable for routine use and is also not available from blood banks. However, there is some evidence in the literature that fresh whole blood transfusion may be helpful in patients with massive trauma as all blood components are replaced simultaneously matching the loss. However, this is difficult practically as the blood products still need to be screened for various antigens and infections.

Other blood products available are fibrinogen concentrate (high risk of hepatitis), Factor VIII and Factor IX concentrates (for use in haemophilia and Christmas disease respectively) and Factor VII concentrate (for use in disseminated intravascular coagulation—DIC).

COMPLICATIONS OF BLOOD TRANSFUSION

I. Immune complications

1. Haemolytic reactions

a. Major (ABO) incompatibility reaction

- This is the result of mismatched blood transfusion.
- Majority of cases are due to technical errors like sampling, labelling, dispatching.
- This causes intravascular haemolysis.

Clinical features

- Haematuria
- Pain in the loins (bilateral)
- Fever with chills and rigors
- Oliguria is due to products of mismatched blood transfusion blocking the renal tubules. It results in acute renal *tubular necrosis.*

Treatment

- Stop the blood. Send it to blood bank and recheck.
- Repeat coagulation profile
- IV fluids, monitor urine output, check urine for Hb
- Diuresis with furosemide 20–40 mg IV or injection mannitol 20% 100 ml IV to flush the kidney.

b. Minor incompatibility reaction

- Occurs due to extravascular haemolysis
- Usually mild, occurs at 2–21 days
- Occurs due to antibodies to minor antigens
- Malaise, jaundice and fever
- Treatment is supportive

2. Nonhaemolytic reactions

a. Febrile reaction

- Occurs due to sensitisation to WBCs or platelets
- Increased temperature—no haemolysis
- Use of 20–40 mm filter or leukocyte-depleted blood avoids it.

b. Allergic reaction

- Occurs due to allergy to plasma products; manifest as chills, rigors and rashes all over.
- They subside with antihistaminics such as chlorpheniramine maleate 10 mg IV.

c. Transfusion related acute lung injury (TRALI)

It is a rare complication, occurring within six hours of a transfusion due to the presence of antileukocyte antibodies in the transfused plasma causing patient's white cells to aggregate in the pulmonary circulation. This leads to degranulation of leukocytes causing increased capillary permeability and noncardiogenic pulmonary oedema. The symptoms may vary from mild dyspnoea to full blown acute respiratory distress syndrome. Proper supportive therapy will see that it resolves within 24–48 hours.

d. Congestive cardiac failure (CCF)

CCF can occur if whole blood is transfused rapidly in patients with chronic anaemia.

Treatment

- Slow transfusion, injection furosemide 20 mg IV
- Packed cell transfusion is the choice in these patients.

II. Infectious complications

Serum hepatitis, *AIDS, malaria, syphilis* are dangerous infectious diseases which can be transmitted from one patient to another through blood transfusion. The danger is increased in cases of multiple transfusions and in emergency situations. ***"Prevention is better than cure"***. Hence, it is mandatory to screen the blood for these diseases before transfusion.

III. Complications of massive transfusion

(Key Box 12.8)

Massive blood transfusion

Definition: Massive blood transfusion has been variously defined—replacement of > 1 blood volume (or > 10 units of packed cells) in 24 hours, half the patient's blood volume in six hours, > 4 RBC units in one hour with ongoing need for transfusion, 500 ml over 5 min or even blood loss > 150 ml/min with haemodynamic instability and need for transfusion. Massive blood loss may occur with trauma, postpartum haemorrhage or during major surgeries.

KEY BOX 12.8

MASSIVE BLOOD TRANSFUSION

1. > 500 ml over 5 minutes
2. > 1/2 the patient's blood volume in 6 hours
3. > the whole blood volume in 24 hours

Problems

- Citrate toxicity—hypocalcaemia
- Thrombocytopaenia
- Clotting factors deficiency
- Acute lung injury

Massive transfusion protocol: In patients with massive bleeding and anticipated to require massive transfusion, the blood bank should be intimated to activate massive transfusion protocol (MTP). A blood sample must be sent for cross matching with an initial request for 4 units of O –ve red blood cells. After this, if MTP is requested, the blood bank releases blood products in different 'boxes' (Table 12.2).

The blood products are obtained box by box as necessary. If the bleeding stops, the blood bank should be intimated immediately so that MTP can be terminated. If bleeding continues, box three and four are alternately requested for. In such patients regular and half hourly measurements of acid-base status, haemoglobin, platelets, prothrombin time (PT), activated partial thromboplastin time (aPTT), fibrinogen and serum calcium are required with an aim to normalise their values.

Disseminated intravascular coagulation (DIC)

- Occurs in massive blood transfusion wherein all factors of coagulation are used up resulting in a bleeding disorder.
- It produces severe afibrinogenemia
- It is treated by replacement with fibrinogen (cryoprecipitate) and other clotting factors.

AUTOLOGOUS TRANSFUSION

This concept originated to avoid transfusion reactions which can develop when homologous blood is used.

- Here, patient's own blood is used.

Types of autologous transfusion

1. Predeposit

- 2–5 units of blood may be donated over 2–3 weeks befor elective surgery.

2. Preoperative haemodilution

- Cases such as surgery for thyrotoxicosis or abdominopelvi resection wherein one can expect 1–2 units of blood los Just before surgery, 1–2 units of blood are removed an retransfused after the procedure.

3. Blood salvage

Blood which was lost during surgery is collected, mixed wit anticoagulant solution, washed and reinfused. This can be don provided surgery does not involve severe infection, bowe resection or malignancy, revision total hip replacement.

Advantage

All the risks involved with blood transfusion are avoided.

Disadvantages

- May not be acceptable to the patient
- Sophisticated equipment required

Blood Product (Fig. 12.1)

PLASMA SUBSTITUTES

These are colloidal solutions used to restore normal blooc volume in emergency situations, e.g. polytrauma with severe haemorrhage, massive GI bleed and shock.

1. Albumin

- It is a rich protein but carries no risk of hepatitis
- It is available as 5 and 20%
- Used in severe burns—acute severe hypoalbuminaemia
- Used in nephrotic syndrome

PEARLS OF WISDOM

Packed cells and fresh frozen plasma transfusion should not be used to treat malnutrition.

2. Gelatins

- Good plasma expander
- Plasma expansion lasts a few hours

Table 12.2 Release of blood products on activation of MTP

Box	PRBC	FFP	Platelet	Cryoprecipitate
One	2	2	–	–
Two	4	4	1 adult	–
Three	4	4	–	3
Four	4	4	1 adult	

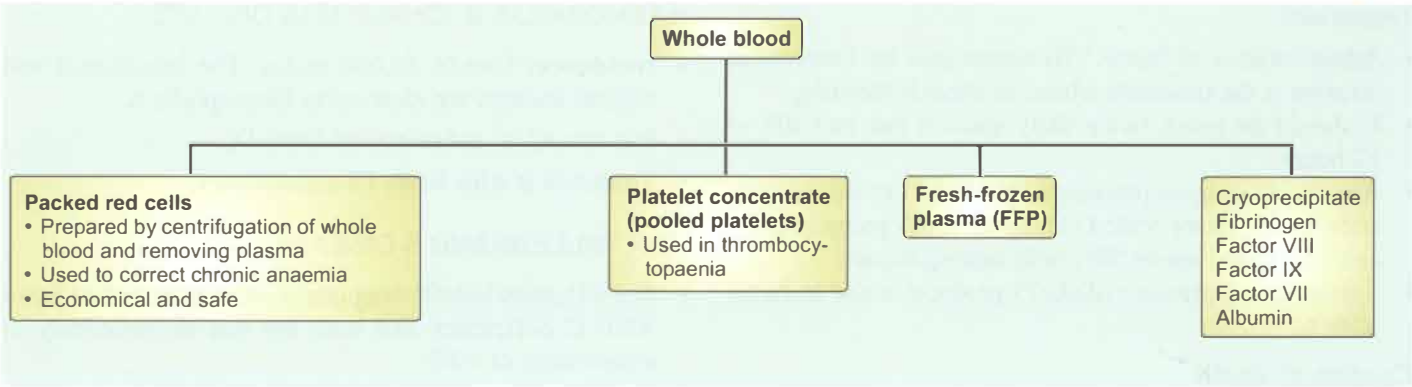


Fig. 12.1: Blood products

- Severe reactions with urea-linked gelatin, e.g. Haemaccel (1:2,000) but less with succinylated gelatin, e.g. gelofusine (1:13,000)

3. Dextran 40
- Reduces viscosity and red cell sludging
- May affect renal function and coagulation

4. Hydroxyethyl starch
- Derived from starch
- Plasma expansion lasts for over 24 hours
- Maximum dose—20 ml/kg
- Large doses may interfere with coagulation
- Incidence of severe reactions (1:16,000).
- Use in shock is associated with increased risk of renal injury and mortality.

BLEEDING DISORDERS

HAEMOPHILIA

This is the commonest bleeding disorder which occurs due to X-linked genetic disorder of coagulation.

Types

1. **Haemophilia A:** This results from a reduction of factor VIII (antihaemophilic factor) and is carried by a recessive gene.
2. **Haemophilia B:** This results due to deficiency of factor IX.

HAEMOPHILIA A

- Haemophilia "excessive bleeding, unusual bleeding, unexpected bleeding" is due to haemophilia 'A'.
- A haemophilic patient's daughters will be carriers but all sons will be normal.
- Thus, a carrier woman has 50% chance of producing haemophiliac male or a female carrier.
- The level of coagulation factor VIII in the blood may be less than 1% of that in a normal individual.

Clinical features

"Excessive bleeding, unusual bleeding, unexpected bleeding" is due to haemophilia 'A' (Table 12.3).

1. Bleeding into joints (haemarthrosis)
- Large joints such as knees, elbows, ankles, wrists are affected.
- Spontaneous bleeding is common. It may also occur due to minor trauma.
- Repeated bleeding may result in permanent damage to the articular surfaces resulting in deformity of the joints.

2. Bleeding into muscles
- Calf muscle and psoas muscle haematomas are common resulting in contraction and fibrosis of muscle, muscle pain and weakness of limb.
- Intramuscular injections should be avoided.

Table 12.3 Blood changes in haemophilia A and von Willebrand's disease

	Haemophilia 'A'	von Willebrand's disease
Bleeding time	Normal	↑
Prothrombin time	Normal	Normal
VIII: C	↓↓↓	↓
vWF	Normal	↓

Treatment

- Administration of factor VIII concentrate by intravenous infusion is the treatment whenever there is bleeding.
- It should be given twice daily since it has half-life of 12 hours.
- Any major surgical procedure should be carried out only after raising factor VIII: C levels to 100% preoperatively and maintained above 50% until healing occurs.
- Synthetic vasopressin (DDAVP) produces a rise in factor VIII: C.

Causes of death

- Cerebral haemorrhage used to be the most common cause of death. However, today HIV infection seems to be the most common cause of death.
- Hepatocellular carcinoma, cirrhosis due to HIV and HCV also are the other causes (due to repeated blood transfusions).

HAEMOPHILIA B (CHRISTMAS DISEASE)

- **Incidence:** One in 30,000 males. The inheritance an clinical features are identical to haemophilia A.
- It is caused by deficiency of factor IX.
- Treatment is with factor IX concentrates.

VON WILLEBRAND'S DISEASE (vWD)

- In vWD, there is defective platelet function as well as facto VIII: C deficiency and both are due to deficiency o abnormality of vWF.
- Epistaxis, menorrhagia and bleeding following mino trauma or surgery is common.
- Haemarthrosis is rare.
- Treatment is similar to mild haemophilia. Thus, DDAVF or IV infusion of factor VIII: C or vWF surgery.

A FEW PHOTOGRAPHS OF PATIENTS WITH SEPTIC SHOCK (Figs 12.2 to 12.7)

Fig. 12.2: Extensive gangrene

Fig. 12.3: Necrotising fasciitis of upper limb

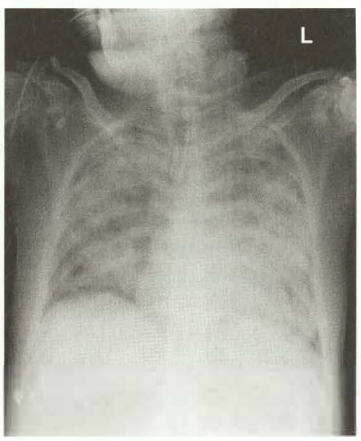


Fig. 12.4: Chest X-ray in septic shock

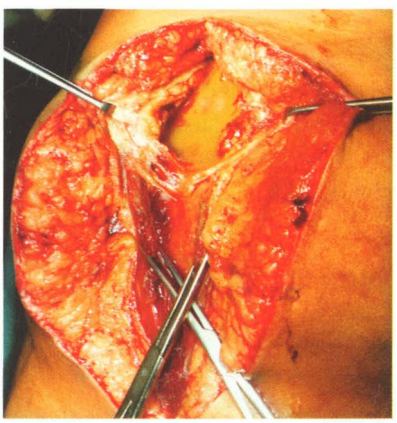


Fig. 12.5: Faecal peritonitis

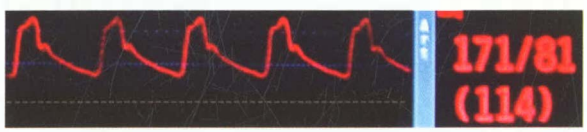


Fig. 12.6A: Intra-arterial blood pressure monitoring

Fig. 12.6B: Pancreatic necrosis—necrosectomy specimen

Fig. 12.7: Septic shock patient recovering

MULTIPLE CHOICE QUESTIONS

1. Which of the following statements is false in management of haemorrhagic shock?
A. Oxygen by face mask in conscious patients
B. Intravenous dextrose is the first agent to be used
C. Legs are elevated
D. Inotropes can also be used in selected patients

2. Which solution is ideal in the initial management of shock?
A. Normal saline B. Ringer lactate
C. Blood D. Albumin

3. Characteristic feature of septic shock include following *except*:
A. Hypotension B. Tachycardia
C. Oliguria D. Alkalosis

4. Following are used to treat early shock *except:*
A. Ringer lactate B. Blood transfusion
C. Isotonic saline D. Albumin

5. Following are uses of hyperbaric oxygen *except:*
A. Carbon monoxide poisoning
B. Gas gangrene
C. Decompression sickness
D. Before chemotherapy

6. In the measurement of central venous pressure, which of the following statements is true:
A. Femoral vein is the route of choice
B. The zero reference point in a supine position is midaxillary line
C. CVP does not change with phases of respiraiton
D. CVP is low in tension pneumothorax.

7. Which one of the following statements is false about central venous pressure?
A. If CVP is high, fluid overload may result in pulmonary oedema
B. CVP is affected by intrathoracic pressure
C. CVP is an indicator of blood volume
D. CVP is almost zero in cardiac tamponade

8. Pulmonary capillary wedge pressure is measured using
A. Foley's catheter
B. Fogarty balloon catheter
C. Swan-Ganz catheter
D. Arterial catheter

9. Which one of the following catheters does not have balloon at the tip?
A. Swan-Ganz B. Foley catheter
C. Fogarty catheter D. Seldinger catheter

10. Packed red cells are used to treat:
A. Chronic anaemia B. Renal failure
C. Tetanus D. Gas gangrene

11. Haemolytic reactions due to mismatched blood transfusion include the following *except*:
A. Haematuria
B. Pain in the loins
C. Fever with chills and rigors
D. Diuresis

12. Following is used to treat disseminated intravascular coagulation *except:*
A. Packed cells B. Platelets
C. Cryoprecipitate D. White blood cells

13. Which one of the following is decreased in disseminated intravascular coagulation?
A. Prothrombin time
B. Partial thromboplastin time
C. Bleeding time
D. Platelets

14. Following are true in haemophilia *except:*
A. Prothrombin time is unaffected
B. Partial thromboplastin time is unaffected
C. Bleeding time is unaffected
D. Platelet count is unaffected

15. Following are true for aspirin *except:*
A. Prothrombin time is unaffected
B. Partial thromboplastin time is unaffected
C. Bleeding time is unaffected
D. Platelet count is unaffected

16. Thrombocytopaenia is seen in following conditions *except:*
A. Disseminated intravascular coagulation
B. Massive blood transfusion
C. Sickle cell anaemia
D. Haemolytic uraemic syndrome

17. Which one of the following drugs does not cause thrombocytopaenia?
A. Heparin
B. Sulfa containing antibiotics
C. Interferons
D. Pencillins

18. Following are true about blood products *except*:
A. Fresh-frozen plasma is given in severe liver failure
B. Cryoprecipitate has not much role in disseminated intravascular coagulation
C. Fibrinogen has high risk of hepatitis
D. Factor VIII is used in haemophilia

19. Following are true about platelet transfusion *except:*
A. Action lasts for 3–5 days
B. Each unit will raise platelet count by 5–10,000 cells/min
C. Platelets must be ABO compatible
D. Platelets can be low after heparin

20. Following are true for albumin *except:*
A. First line of treatment in hypovolaemic shock
B. Does not carry risk of transfusion hepatitis
C. Used in nephrotic syndrome
D. It should not be used to treat malnutrition

21. In haemorrhagic shock, when both the systolic and diastolic blood pressures are normal, the patient is said to be in Class ____ shock.
A. IV B. I
C. II D. III

22. In haemorrhagic shock, when the patient has lost 30–40% blood volume, he is said to be in Class ____ shock.
A. I B. II
C. III D. IV

23. Tension pneumothorax is one of the reasons for which of the following types of shock?
A. Hypovolaemic shock
B. Obstructive shock
C. Distributive shock
D. Septic shock

24. The drug of choice in anaphylactic shock is:
A. Histamine
B. Adrenaline
C. Promethazine
D. Chlorpheniramine

25. The 'zero reference point' for arterial pressur transducers in patients in supine position should b at:
A. Level of nipple
B. Manubriosternal junction
C. Xiphoid process level
D. Radial arterial level.

26. The central venous pressure is high in the following type of shock:
A. Hypovolaemic shock
B. Anaphylactic shock
C. Septic shock
D. Obstructive shock

27. The following are the causes of obstructive shock *except:*
A. Chest injury
B. Pulmonary embolus
C. Cardiac tamponade
D. Liver laceration

28. The following colloid is often used to reduce plasma viscosity:
A. Dextran 40 B. Hetastarch
C. Haemaccel D. Albumin

29. The route of choice for administration of adrenaline in anaphylactic shock is:
A. Intramuscular B. Intravenous
C. Subcutaneous D. Inhalational

30. The blood product of choice in bleeding patients with a fibrinogen concentration of < 100 mg/L:
A. Packed cells
B. Fresh-frozen plasma
C. Platelets
D. Cryoprecipitate

ANSWERS

1 B	2 B	3 D	4 D	5 D	6 B	7 D	8 C	9 D	10 A
11 D	12 D	13 D	14 B	15 C	16 C	17 D	18 B	19 C	20 A
21 B	22 C	23 B	24 B	25 B	26 D	27 D	28 A	29 B	30 D

13

Burns, Skin Grafting and Flaps

- Burns
- Free skin grafting
- Flaps
- Electrical burns
- Chemical burns
- Hydrotherapy
- What is new?/Recent advances

BURNS

Definition

Burns is a type of coagulative necrosis caused by heat, transferred from the source to the body.

Frostbite which occurs in cold countries is also a coagulative necrosis but it is caused by extreme degrees of cold. Scald is a burn caused by moist heat (steam). Burns never occur at temperatures less than 44°C.

Types

- Thermal: Flame burns and scald burns
- Electrical
- Chemical
- Radiation

Pathophysiology of burns shock

Although the exact mechanism of the postburn microvascular changes and hypovolaemia leading to low cardiac output and poor tissue perfusion has not been determined, the following mechanisms have been proposed:

1. **Increased capillary permeability** leading to fluid and protein leakage from the intravascular space.
2. **Decreased plasma oncotic pressure** due to hypoproteinaemia resulting from loss of protein from the intravascular space.
3. **Increased capillary hydrostatic pressure** due to vasoconstriction or partial blockage of vessels with aggregate of cells and platelets.
4. **Reduced clearance of fluid and protein** from the interstitial space by lymphatic ducts due to blockage by platelet aggregates and fibrin clots.
5. **Intracellular fluid accumulation** due to impaired cell membrane function.
6. **Increase in osmotic pressure in the burned tissue** leading to further fluid accumulation.
7. **Increased evaporative water loss** (Key Box 13.1)
8. **Depressed myocardial function**
   - Chemical mediators released from the site of injury are responsible for the development of typical inflammatory response. This results in rapid and dramatic oedema formation.
   - The activated complement cascade system facilitates liberation of various permeability factors such as histamine, prostaglandins (PGF-1, PGF-2, PGF-2α) and thromboxane.

KEY BOX 13.1

BURNS PATIENT AND THE KIDNEY

- Maximum water reabsorption by release of ADH from posterior pituitary, as a compensatory mechanism.
- Aldosterone is released to cause maximum sodium reabsorption.
- Acute tubular necrosis (ATN) can occur due to toxins.
- Injury to kidney can occur due to myoglobin.

- Macromolecular leakage into burned areas, catabolism and reduced immunoglobulin synthesis results in a decreased concentration of all individual immunoglobulin levels and triggering of complement cascade.
- **Inhalation injury:** Glottic oedema, necrotising bronchitis, pneumonia are the dangerous events that follow an inhalation injury (Key Box 13.2).
- **Septic shock:** Due to infection by micro-organisms. Infection commonly occurs from cross infection. GI system, genitourinary system, respiratory system, IV cannula site are various other portals of entry of infection (Fig. 13.1).
- **Endotoxins:** Lipopolysaccharides and endogenous chemical mediators (cytokines) such as TNF-α and interleukins are activated resulting in systemic inflammatory response syndrome and multiorgan failure.

KEY BOX 13.2

AIRWAY oedema in burns

- It is due to inhalation injury
- Direct injury from hot air/smoke
- Inflammatory response to burns
- Oedema from resuscitation fluids
- Suspect in patients with facial burns, tachypnoea, and progressive hoarseness
- Oxygen, pulse oximetry
- Bronchoscopy, early endotracheal intubation

Characteristic Ebb and flow of burns

- **Ebb:** Low metabolism and temperature; cardiac output i decreased.
- **Flow:** Hypermetabolism, high cardiac output, hyper glycaemia, increased heat production.

Metabolic response to burn injury

- **Gluconeogenesis** and glycogenolysis is increased.
- **Lipolysis:** It is increased and fatty acids are released. They are re-esterified into triglycerides resulting in fa accumulation in the liver.
- **Proteolysis:** The result is increased production of urea which is excreted in the urine. As a result of this, there is increased efflux of amino acids from skeletal muscle poo including alanine and glutamine.
  Significance: Burns patient requires more than 1 g/kg/day proteins.
- Glutamine can also be given
- **Catecholamines** are massively elevated in burn injury.

Emergency care

- Any patient exposed to smoky fire should receive 100% oxygen *via* a non-rebreathing mask. If he is unconscious, endotracheal intubation should be performed.
- Intravenous line and administration of lactated Ringer solution at 1 L/h in adult.
- Transport the patient-warm and wrapped in a clean sheet. Clothing and jewellery should be removed because the swelling begins immediately.

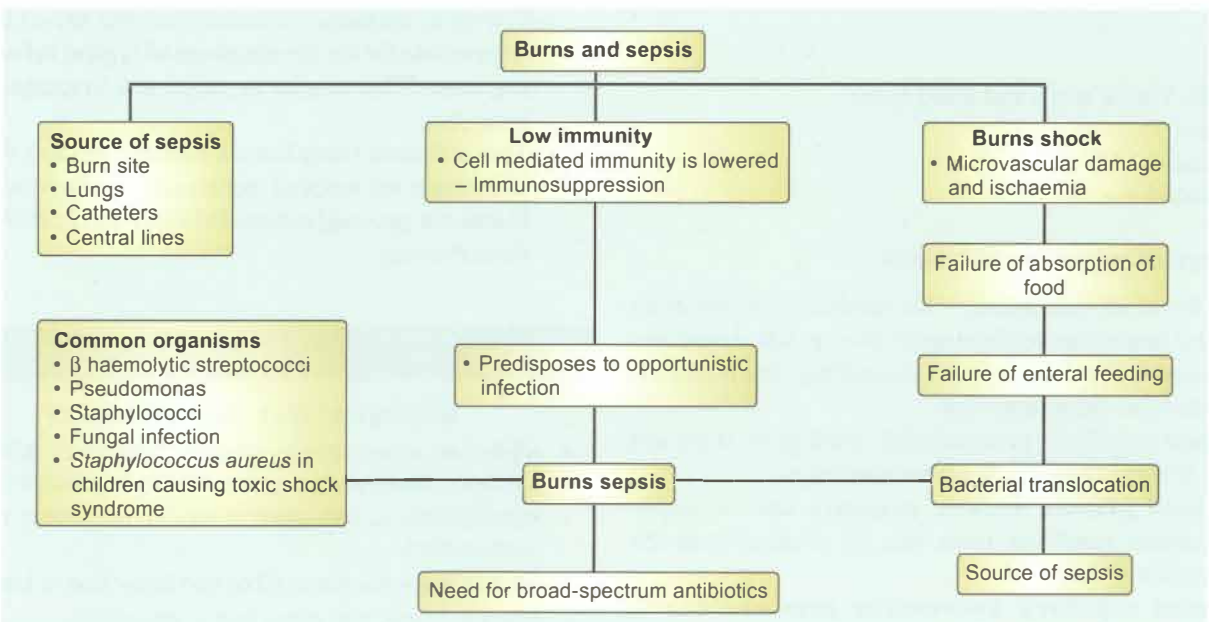


Fig. 13.1: Burns and sepsis

- Small burns can be managed by cool water. **Do not use ice-cold water.**

Management of burns patients

1. **First aid:** Cold water bath should be given immediately. This takes away the heat, stabilises mast cell, thus decreasing the release of histamine and reduces oedema.
2. Careful history-taking AMPLE (Key Box 13.3). **The time since sustaining the injury,** the type of heat source, location and circumstances of the burn should be recorded.
3. Hospitalisation and admission in a burns ward with air-conditioning facility.
4. **Assessment of depth of burn** (Table 13.1)
   - **Partial thickness burns:** Here, superficial layers of skin are destroyed. Epidermis and variable portion of dermis are involved. Since the nerve endings are exposed, it causes severe degree of pain.
   - **Full thickness burns:** Involvement of full thickness of dermis with epidermis. Since, the nerves are destroyed, it is less painful.
5. **Assessment of extent of burns in terms of body surface area (BSA):** It is calculated by a ***Rule of 9 "Rule of Wallace"***
   - Burns of head and neck: 9%
   - Burns of upper limbs: 9 × 2 = 18%
   - Burns of anterior trunk = 18%
   - Burns of posterior trunk = 18%
   - Burns of the lower limbs: 18 × 2 = 36% (front and back of each lower limb is 9%)
   - Burns of external genitalia: 1%
6. Temperature, pulse, respiration and blood pressure are monitored and maintained within normal limits.
7. An indwelling **urinary catheter** (Foley catheter) is introduced, and strict intake and output chart must be maintained.
8. A **Ryle's tube** is passed. A burn patient can develop "acute stress ulcers" called ***acute peptic ulcers*** or ***Curling ulcers***. Hence, to prevent bleeding, a cold stomach wash is given through the Ryle's tube. Antacids and H_2 receptor blockers such as Ranitidine 150 mg twice a day are also given.

KEY BOX 13.3

AMPLE (Mnemonic)

Allergy
Medications
Past medical history
Last meal
Events regarding injury

9. **Replacement of fluid volume is done using anyone of the following formulas**

   a. Muir and Barclay formula

   $$1 \text{ ration} = \frac{\% \text{ of burns} \times \text{body weight (kg)}}{2}$$

   3 rations in 12 hours, 2 rations in next 12 hours and 1 ration in next 12 hours.

   Example: 40% burns patient weighing 60 kg.

   $$1 \text{ ration} = \frac{40 \times 60}{2} = 1200 \text{ ml}$$

   1st 12 hours : 3600 ml fluid

   2nd 12 hours : 2400 ml fluid

   3rd 12 hours : 1200 ml fluid

- The best solution for replacement is plasma. For fear of transfusion reactions, a crystalloid such as ***Ringer lactate*** can be used.
- In any patient with burns above 20%, packed red cells are given in the 2nd ration (after 12 hours).

PEARLS OF WISDOM

Loss of intravascular fluid will start when burns size approaches 10–15% of total body surface area (TBSA). When TBSA is 25%, loss of fluid will start from vessels in the remote area also.

- After 36 hours, depending upon the urinary output and depending on the requirements of the body, about 2–3 L of fluid are given per day.

   b. Parkland's formula: 4 ml/%/kg in the first 24 hours. 1/2: In 1st 8 hours, 1/4: Next 8 hours each.

Table 13.1 Summary of three types of burns

	Involved area	Pain	Adnexa	Appearance	Healing
I° Burn	Only epidermis	+	+	Erythema/oedema	3–5 days without scar
II° Burn	Varying depth of dermis	++/–	+	Blister, soft waxy, white, elastic, pain sensation to needle prick present	10–20 days, with hypertrophic scar
III° Burn	Involvement of entire depth of epidermis and dermis	– –	Lost	Tough, dry, eschar, thrombosed subcutaneous vein. Pain sensation to needle prick absent	3–5 weeks, eschar separates. **Always needs** split skin graft

c. Modified Brooke formula

1st 24 hours: 2 ml/% of burns/kg (deficit) + maintenance (2500 ml).

2nd 24 hours: 50% of the deficit in 1st 24 hours + maintenance (2500 ml in adults).

Rate of administration: 50% in the first 8 hours after injury and 50% in the next 16 hours.

10. **Broad-spectrum antibiotics** are given against gram +ve, gram –ve and anaerobic organisms.
    - To treat septic shock, higher antibiotics such as cephalosporins have to be given.

PEARLS OF WISDOM

DO NOT GIVE COLLOID in first 24 hours.

11. Narcotic analgesics such as injection morphine 10–12 mg **IM** or injection pethidine 60–75 mg **IM** is given to relieve the pain. (If the veins are not visible, a "cut down" needs to be done, so that the infusion cannula can be maintained for a longer time.)

Treatment of burns wound

1. Clean the wound with antiseptic agents such as savlon, iodine.
2. **Dressing**

   Open method: Exposure line of management: If facilities available are good, with a fumigated ward, the wound can be left open to the atmosphere after applying *Silver Sulfadiazine* cream.

   Closed method: Alternately, after applying silver sulfadiazine, 1% silver nitrate and mefenide cotton rolls are applied, over which a bandage is applied. This is called closed method. Advantages of closed method are less pain, reduced infection and sogging, and better medication.
3. **Surgery: Tangential excision** followed by grafting is done after about 48–72 hours in which the burn wound is excised *tangentially* till fresh bleeding occurs. This is followed by skin grafting. Skin is taken from the remaining normal area, or can be obtained from skin bank. A mesh expander and cell culture may also be used.
4. Treatment of circumferential deep 2nd and 3rd degree burns.

Advantages of tangential excision

1. It prevents secondary infection and septicaemia.
2. Decreases the hospital stay (2–5 weeks is the usual time taken for eschar of III° burn to separate. This is reduced by tangential excision).
3. Decrease of incidence of contractures and hypertrophic sca
4. Cost of the treatment is reduced.

Causes of death in burns patient

1. Uncontrolled hypovolaemic shock (refractory hyper volaemia).
2. Choking, suffocation due to respiratory burns especially i burns of head and neck.
3. *Septicaemic shock* is the most common cause of death.
   Mnemonic: Look for 6 **P**s
   Pain, **P**allor, **P**ulselessness, **P**araesthesia, **P**aralysis **P**oikilothermia (in the affected part).
   - If these are present, it indicates compartment syndrom
   - It needs to be treated by escharotomy.

Prognosis

- It depends upon several factors (Key Box 13.4), the mos important being percentage of burns and facilities o treatment.
- If untreated, postburns contractures can occur (Figs 13.2 to 13.4).

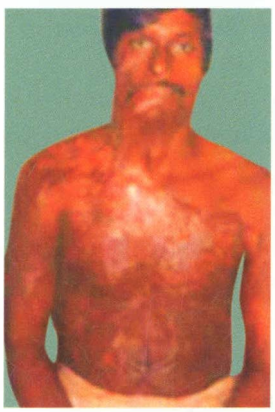


Fig. 13.2: Postburn contracture in an adult

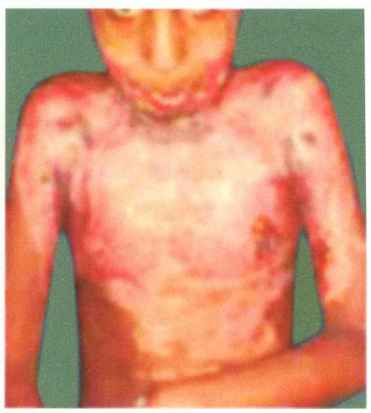


Fig. 13.3: Postburn neck contracture in a child

Fig. 13.4: Hypertrophic scar in a deep dermal burns

KEY BOX 13.4

FACTORS AFFECTING OUTCOME

- Extremes of age
- Depth of burns
- Inhalation of noxious fumes
- Cardiopulmonary disease

FREE SKIN GRAFTING

Skin grafting is the commonest method of achieving wound cover.

Types

1. Split skin graft (SSG—Thiersch graft)

- Also called partial thickness graft
- Consists of epidermis and a variable portion of dermis (Figs 13.5 and 13.6)
- Split skin graft is usually harvested using **Humby's knife** (Fig. 13.7). Drum dermatome or a power dermatome may also be used.
- Preferred donor area is thigh

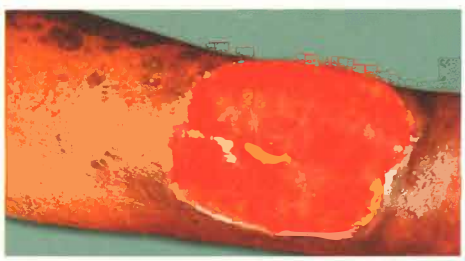


Fig. 13.5: Wound is ready for skin grafting. Observe the red granulation tissue

Fig. 13.6: Skin graft is applied over the recipient area

Fig. 13.7: Skin graft is taken from thigh using Humby's knife

2. Full thickness graft (Wolfe graft)

- Consists of epidermis and full thickness of dermis.
- Harvested using ordinary scalpel.
- Needs excellent vascularity of the recipient wound for graft survival.
- Used for small uncontaminated wounds produced after excision of skin lesions or after release of skin contractures (lower eyelids, fingers).
- **Donor area needs primary suturing or split skin graft** for healing and hence limits the size of the graft.
- Unlike split skin grafts, full thickness grafts do not contract and retain their colour. Hence, they are cosmetically superior.

Indications of skin graft (Key Box 13.5)

1. Skin loss

- **Post-traumatic** (e.g. avulsion and degloving injury)
- **Post-surgical** (e.g. excision of tumours, excision of burn wound)
- As a result of **pathological process** (e.g. venous ulcer, diabetic ulcer)

2. Mucosa loss

- After **excision** of lesions of oral cavity, tongue.
- For **resurfacing** reconstructed vagina in cases of vaginal agenesis.

Contraindications for skin graft

- Infection by ***beta-haemolytic streptococci.*** They produce fibrinolysin which dissolves fibrin.
- Presence of an **infected wound** with copious discharge in the vicinity.
- **Avascular wounds:** With exposed bare bone without periosteum, exposed tendon without **paratenon** and exposed cartilage without perichondrium.

Healing of the donor area

Donor area of split skin graft heals by epithelialisation from the adnexal remnants of dermis, pilosebaceous follicles and/or sweat gland apparatus. Complete healing of donor area occurs by 8–10 days.

KEY BOX 13.5

IDEAL REQUIREMENTS FOR FREE SKIN GRAFT

1. Wound should be free from infections such as Streptococci and Pseudomonas
2. Vascular wounds, e.g. wounds with healthy granulation.
3. Wound should be thoroughly debrided.
4. Haemostasis must be achieved before placing the graft.
5. Close and immobile contact between graft and the wound.
6. Recipient area should be immobilised with **POP** slab.

The process of graft 'Take'

- The processes which result in **reattachment** and **revascularisation** of the graft to the bed are collectively referred to as **"take" of graft**.
- The graft initially adheres to its new bed by fibrin. Revascularisation starts by 48 hours and is completed by 4–5 days. This is achieved by the outgrowth of capillary buds from the recipient area to unite with those on the deep surface of graft. For the first 2 days after grafting, the skin graft derives its nutrition from the wound by the process of serum imbibition/plasmatic circulation.

Fig. 13.8: PMMC flap following radical parotidectomy

Fig. 13.9: Latissimus dorsi flap used to cover the defect in the arm following radical excision of a sarcoma

FLAPS

Flap is a block of tissue transferred from donor to recipient area along with its vascularity.

Common indications for flap surgery

- To cover defects/wounds **where free skin graft cannot be used, e.g. exposed bare bones, bare tendons, bare cartilage.**
- Wounds with exposed joints, exposed major vessels and nerves.
- **Implant exposure** following orthopaedic procedures.
- In wounds with **soft tissue loss**, where future reconstructive surgery is contemplated.
- Defects which need better contour to improve cosmesis.
- Breast reconstruction following mastectomy.

Classification of flaps

These are broadly classified into Pedicled flaps and Free flaps.

I. Pedicled flaps

Pedicle or the base remains attached to the donor site during its transfer to the recipient area (Figs 13.8 to 13.10). Pedicled flaps may be of following types:

1. **Local flaps**, e.g. rotation, transposition, limberg and bilobed.
2. **Pectoralis major myocutaneous (PMMC)** flap, deltopectoral (DP) flap for head and neck defects, transverse abdominis myocutaneous (TRAM) flap for breast reconstruction.

   Regional flap, e.g. PMMC, DP for head and neck defects, TRAM for breast reconstruction.
3. **Distant flaps**, e.g. groin flap, subaxillary flap for hand defects.

Fig. 13.10: Bedsore covered with local advancement flap

A few examples of the pedicled flap

- Skin flap
- Fasciocutaneous flap
- Muscle flap
- Myocutaneous flap
- Adipofascial flap
- Osteocutaneous flap

II. Free flaps

These are completely detached from the donor area before being transferred to the recipient area. The vascularity of the flap at the recipient site is immediately restored by anastomosing the vessels of the flap with the vessels at the recipient area using microvascular techniques.

Some of the commonly performed flaps

- **Forehead flap:** Entire forehead skin can be raised based on anterior branch of superficial temporal artery. It bears an unsightly scar of the donor site. **Median (Indian) forehead flap** based on supratrochlear vessels is a very useful flap in reconstructing defects over nose.

- **Deltopectoral (DP) flap:** It is supplied by upper 4 perforating branches of internal mammary artery and is used to reconstruct defects of neck and lower face. After about 4 weeks, the flap is divided and the base is returned to the chest wall.
- **Pectoralis major myocutaneous (PMMC) flap:** Pectoral branch of thoracoacromial artery is the pedicle of this flap. It is the ideal pedicled flap for reconstruction of head and neck defects following ablative surgeries for various head and neck cancers. Hence, it is described as a workhorse among the flaps. Osteomyocutaneous PMMC **flap by including 5th or 6th rib can be used for mandibular reconstruction.**
- **Latissimus dorsi flap (LDF):** As a myocutaneous flap, based on thoracodorsal vessels can be used for reconstruction of the lower half of face, neck, breast, chest wall, axilla, and upper arm.

Free (Microvascular) flaps

Using operating microscope and microvascular techniques, it is possible to do a free tissue transfer of tissues such as skin, muscle, bone, intestine, omentum, etc. The procedure involves anastomosis of vessels of the flap to vessels at the donor site, e.g. latissimus dorsi muscle myocutaneous flap, radical artery forearm flap, gracilis flap and free fibula flap.

ELECTRICAL BURNS

The most important factors taken into consideration while assessing the damage caused by the passage of electrical current through the body are: **The nature of electric current and the resistance of body tissues through which the electric current is passing.**

- When a portion of the body comes in contact with a live electric wire, the actual point where the electric current enters the body is known as point of entry and where it is grounded as the point of exit. The most important factor which decides the severity of injury is the **"voltage"** of current.
- The resistance offered by the tissues to the flow of current leads to the **conversion of electrical energy to thermal injury** and causes tissue damage.

Types of injuries

- **True electrical injury:** The burns is as a result of heat generated due to the passage of current.
- **Flame burns:** It is due to an electrical flash or spark.
- **Arc burns:** Localised injury due to intense heat at the termination of current flow. It occurs when it jumps the gap between the source and the conductor, e.g. flexor aspect of joints.

Classification

A. Acute injuries

I. Burn injuries due to electric current itself

a. Low voltage injuries (less than 1000 volts of current)
(i) Flash burns (ii) Contact burn.

b. High voltage injuries (more than 1000 volts of current)
(i) Flash (ii) Arc or contact
1. Punctate
2. Extensive
3. Extensive with vascular impairment of extremities.
   - Compartment syndrome and dry gangrene

II. Other associated injuries

a. Thermal burns due to ignited cloth
b. Acute central nervous system complications such as transient loss of consciousness, agitation, and confusion. Rarely, hemiplegia, aplasia, convulsion and memory defects can occur.
c. Acute peripheral neuropathy
d. Cardiac/respiratory arrest
e. Injuries to other internal organs due to fall or electric current.

B. Delayed injuries

1. Delayed spinal cord injury
2. Delayed peripheral neuropathy
3. Optic nerve atrophy, cataract

Summary (Key Box 13.6)

KEY BOX 13.6

SUMMARY OF MANAGEMENT OF ELECTRICAL INJURY

- Initial resuscitation as in thermal burns
- Low voltage injury
  Superficial—conservative
  Deep dermal—tangential excision and split skin graft
  Contact burns—excision with SSG/flap
- **High voltage injury**
  Flash burns: Superficial—conservative
  Deep dermal: Tangential excision and split skin graft
- **Arc or contact burns:** Vascular compromise—immediate fasciotomy, amputation
- **Punctate wounds:** Excision and grafting
- **Extensive injury without vascular compromise:** Wider excision with split skin graft or flap cover
- Recipient area should be immobilised with **POP** slab

CHEMICAL BURNS

- Chemicals are a relatively uncommon cause of burns. The chemicals used in industry, science laboratories and at home are the usual agents.

- The tissue damage in chemical burns is mainly due to prolonged contact period and effects of systemic absorption.

Classification of agents that cause injury

1. **Acids** : Hydrochloric acid, sulphuric acid, nitric acid, hydrofluoric acid, phenol (carbolic acid), oxalic acid
2. **Alkalis** : Sodium hydroxide, potassium hydroxide, ammonium hydroxide, lithium, barium and calcium hydroxide
3. **Others** : **Inorganic substances**
   Phosphorus, wet cement lime, potassium permanganate
   Organic substances
   Kerosene, petrol, turpentine, naphthalene.

Modes of action of chemicals on tissues

- **Acid** causes coagulative necrosis of the skin due to rapid conversion of protein to coagulum salt of the acid. The coagulated eschar prevents these acids to penetrate deeply. Activity in the tissues continues for a long time.
- **Alkalis** are corrosive agents and produce extensive denaturation of tissue proteins. These produce more tissue destruction than acids.

First aid

- **Early irrigation** of injured area with large volume of water or running water (with few exceptions) is the main focus in the first aid management of chemical injuries. Hydrotherapy mechanically cleans the area reducing the concentration of chemicals and duration of contact. Earlier the hydrotherapy is started, more is the benefit obtained.
- Since the absorption of phenol increases with dilution, surface phenol is removed by solvent **polyethylene glycol before hydrotherapy** is started.

Definitive wound closure

Achieved in a similar way as in any other thermal injury

- **Primary excision and split skin grafting,** of all acid and alkali burns, if done before 10 days.
- **Grafting of granulating wound**, if the patient presents late, with wound infection. This takes much longer (1½ months) to heal compared to thermal burns (3 weeks).

Ocular injuries

Very common in acid and alkali burns. Severe blepharospasm and forceful rubbing increases the severity of injury. Various sequences of events are:

- Sloughing of corneal epithelium
- Stromal oedema
- Corneal ulceration
- Perforation
- Panophthalmitis
- If perforation does not occur, corneal opacity and heavy vascularisation occurs. Quick, thorough and prolonged lavage of cornea with water is the most important measure in the first aid treatment. Systemic and topical steroids minimises inflammation and scarring of cornea.

MISCELLANEOUS

Skin substitutes

- This is one of the important requirements wherein large surface area of burns and skin is lost.
- An ideal substitute must be affordable, permanent, provide normal pigmentation, resist scar formation and grow with developing children.
- A few examples of skin substitutes are:
  - **Dermal substitutes:** They allow for creation of a 'neodermis'. They are formed from patient's own mesenchymal cells. Once 'neo-dermis' is formed, split thickness skin graft is applied. Thus, the burns site is closed quickly with less scarring.
  - **Cultured epithelial autograft** is another example of dermal substitute. They are cultured from patient's own full thickness biopsy. It will require 3 weeks to grow.
  - Another cultured skin is a **biologic dressing** from cultured **neonatal keratinocytes** and **fibroblasts.** They are all very expensive.

Hydrotherapy in burns

It refers to usage of external water (may be very cold) to run over the burned part for a few minutes.

Methods used are:

1. Immersion hydrotherapy. It is performed in tubs called hydrotanks or burns tanks. Tanks should be disinfected after usage. Water used is sterile.
2. Shower hydrotherapy: Depending upon severity of burns, the shower can be either in supine or sitting position. Showering immediately rinses away dead skin and bacteria.

How does it work?

- Cleans the surface of wound and remove debris
- Removes pus
- Prevents loss of fluid through skin
- Minimises scar formation
- Provides moist environment for wound healing
- Minimises risk of infection

Duration

- It may vary from 10 to 20 minutes

Sedation

- Sedation or general anaesthesia is required as it can be very painful.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- The text is updated
- Hydrotherapy is added

MULTIPLE CHOICE QUESTIONS

1. Following factors contribute to burns shock *except:*
A. Increased capillary permeability
B. Increased plasma oncotic pressure
C. Increased capillary hydrostatic pressure
D. Depressed myocardial function

2. Bacterial translocation can occur in the following conditions *except:*
A. Burns
B. Subacute bacterial endocarditis
C. Intestinal obstruction
D. Myocardial infarction

3. Acute tubular necrosis can occur in burns due to the following factors *except:*
A. Hypovolaemia B. Toxins
C. Myoglobin D. Aldosterone

4. Following metabolic response to burns occur *except:*
A. Gluconeogenesis is increased
B. Lipolysis is increased
C. Catecholamines are increased
D. Proteolysis is decreased

5. Tangential excision refers to:
A. Excision and leaving the wound open immediately after admission
B. Excision within 6 hours and primary closure
C. Excision after 2–3 days and skin grafting
D. Excision after 10 days

6. Following are true for split skin graft *except:*
A. It is partial thickness graft
B. It is called Thiersch graft
C. Humby's knife is used often
D. Cosmetically it is superior to full thickness graft

7. Following are true for full thickness graft *except:*
A. It is called Wolfe graft
B. It consists of epidermis and dermis
C. Cosmetically it is superior to partial thickness graft
D. Problem with this graft is that it contracts often

8. Following heals by serum imbibition process:
A. Split skin graft
B. Musculocutaneous flaps
C. Fracture of bones
D. Mesh healing

9. Following are true for pectoralis major myocutaneous flap *except:*
A. It is the flap for head and neck reconstruction
B. Based on pectoral branch of thoracoacromial artery
C. It is an example of pedicle flap
D. It is a free flap

10. Following is true for hydrotherapy in burns *except:*
A. It can be done by immersion
B. Shower can also be used
C. It is painless
D. It rinses away dead skin and bacteria

ANSWERS

1 B	2 D	3 D	4 D	5 C	6 D	7 D	8 A	9 D	10 C

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14

Acid–Base Balance, Fluid and Electrolytes

- Basic definitions
- Henderson-Hasselbalch equation
- Regulation of acid–base balance
- Acid–base disorders
- Rapid interpretation of an ABG report
- Normal physiology
- Water regulation
- Disturbances of volume
- Regulation of sodium concentration
- Disturbances in concentration
- Disturbances in composition of body fluids
- Clinical notes
- Perioperative fluid therapy
- Special purpose solutions
- Nutrition

Introduction

Human blood has a hydrogen ion concentration [H^+] of 35 to 45 nmol/L and it is essential that its concentration is maintained within this narrow range. Hydrogen ions are nothing but protons which can bind to proteins and alter their characteristics. All the enzymes present in the body are proteins and an alteration in these enzyme systems can change the homeostatic mechanisms of the body. Hence, a disturbance in acid–base balance can result in malfunction of the various organ systems.

BASIC DEFINITIONS

What is pH?

pH notation is a more common method of expressing the hydrogen ion concentration. It is defined as the negative logarithm to base 10 of the [H^+] expressed in mol/L. pH of blood = 7.4

What is an acid? What is a base? What is a buffer?

- An **acid** is a substance that dissociates in water to produce H^+.
- A **base** is a substance that accepts H^+.
- A **buffer** is a combination of a weak acid and its conjugate base. By combining with a strong acid or a strong base, they produce the corresponding salt and a weak acid or a weak base respectively.

For example: A weak acid such as carbonic acid with its conjugate base, sodium bicarbonate is called the bicarbonate/carbonic acid buffer system. When a strong acid such as hydrochloric acid is added to the solution, it combines with the weak alkali (sodium bicarbonate) to form sodium chloride and carbonic acid. When a strong base such as sodium hydroxide is added, it combines with the weak acid (carbonic acid) to form sodium carbonate and water. Thus, a strong acid and a strong base are converted into a weak acid and a weak base by the bicarbonate/carbonic acid buffer system.

The hydrogen ion concentration of blood is maintained within narrow limits because of the presence of buffers in the body. **These natural buffers are of two types: Extracellular and intracellular.**

- The extracellular buffers are bicarbonate/carbonic acid buffer system, phosphate buffer system and plasma proteins. The intracellular buffers are haemoglobin and other proteins.
- The most important buffer system in the body is the bicarbonate—carbonic acid buffer system. This is because of the ability of the body to maintain or alter the concentrations of its two components separately. The concentration of carbonic acid is regulated by respiration wherein the excess carbonic acid is eliminated as carbon dioxide by the lungs. The bicarbonate concentration is independently regulated by the kidneys.

THE HENDERSON AND HENDERSON-HASSELBALCH EQUATIONS

The hydrogen ion concentration is proportional to the concentration of buffer systems of the body. The hydrogen ion concentration, carbonic acid levels and the bicarbonate levels of blood are related according to the following equation:

$$[H^+]\ (nmol/L) = K \times \frac{H_2CO_3\ (mmol/L)}{HCO_3^-\ (mmol/L)}$$

Where K = constant. This equation is called the Henderson equation.

The amount of carbonic acid in the blood is directly proportional to the partial pressure of carbon dioxide in the blood. Thus, the carbonic acid concentration is a product of the partial pressure of carbon dioxide in blood times its solubility coefficient.

$[H_2CO_3] = \alpha\ PCO_2$, where α = solubility coefficient of carbon dioxide in blood and PCO_2 is the partial pressure of carbon dioxide in blood.

α = 0.03 ml/mmHg/100 ml blood and normal PCO_2 = 40 mmHg

$[H_2CO_3] = \alpha\ PCO_2 = 0.03 \times 40 = 1.2$ ml/dl.

K = 800 for the carbonic acid/bicarbonate buffer system. The normal bicarbonate level of blood is 24 mmol/L.

$$[H^+]\ (nmol/L) = \frac{800 \times 1.2}{24} = 40\ nmol/L$$

The Henderson equation can also be written as follows:

$$[H^+]\ (nmol/L) = K \times \frac{\alpha\ PCO_2\ (mmHg)}{HCO_3^-\ (mmol/L)}$$

From this equation, it is evident that the hydrogen ion concentration increases when the PCO_2 increases or when the $[HCO_3^-]$ levels decrease. Similarly, a decrease in hydrogen ion concentration occurs when the PCO_2 decreases or when the $[HCO_3^-]$ levels increase.

When expressed in logarithmic form, the Henderson equation is written as follows:

$$pH = pK_a + \log \frac{[HCO_3^-]}{[H_2CO_3]}$$

This logarithmic version of Henderson equation is called the Henderson-Hasselbalch equation.

The pK_a (negative logarithm of the constant K) of the carbonic acid/bicarbonate buffer system is 6.1.

$$pH = 6.1 + \log 24/1.2$$
$$= 6.1 + \log 20 = 6.1 + 1.3 = 7.4$$

It is important to appreciate that the $[H^+]$ and pH are inversely related. When the $[H^+]$ rises, the pH decreases and vice versa.

REGULATION OF ACID–BASE BALANCE

The normal pH of blood is 7.35–7.45. Acidosis is defined as a pH less than 7.35. Conversely, when the pH is more than 7.45, alkalosis is said to exist. Acidosis and alkalosis are of two types each: respiratory and metabolic.

An increase in carbon dioxide (CO_2) levels increases the plasma $[H^+]$ and decreases the pH (respiratory acidosis). Similarly, a decrease in plasma carbon dioxide levels reduces the $[H^+]$ and increases the pH (respiratory alkalosis). A decrease in $[HCO_3^-]$ reduces the pH and is called metabolic acidosis. Similarly, an increase in $[HCO_3^-]$ increases the pH and produces metabolic alkalosis.

The pH is regulated in the human body mainly by two organs: the **respiratory system** and the **renal system**.

The arterial carbon dioxide levels are regulated by the respiratory system. Any increase in carbon dioxide levels stimulates the respiratory centre in the medulla thus augmenting respiration, alveolar ventilation and elimination of extra CO_2 levels. A decrease in CO_2 levels may reduce the stimulus to breathe and cause hypoventilation. This response is limited by hypoxia as the hypoxic drive stimulates the patient to maintain respiration. Respiratory response to changes in CO_2 level occurs very fast.

The plasma bicarbonate levels are regulated by the kidneys. Any decrease in $[HCO_3^-]$ stimulates the kidney to retain and synthesise bicarbonate. High $[HCO_3^-]$ results in elimination of more bicarbonate in urine. In general, the pulmonary response to a change in acid–base status is faster and occurs immediately. However, renal regulation takes time, a few hours to days. Kidneys filter and reabsorb all the bicarbonate in the urine. When necessary, kidneys can also produce extra bicarbonate through the glutamine pathway.

ACID–BASE DISORDERS (Key Box 14.1)

When an acid–base disorder occurs, the initial disturbance that occurs is termed the primary disorder. The body attempts to normalise the pH by certain compensatory mechanisms resulting in a secondary disorder, e.g. primary metabolic acidosis results in an increase in hydrogen ions and a consequent decrease in bicarbonate ions. To compensate for this, the patient hyperventilates and reduces the arterial carbon dioxide levels, thus moving the pH back to normal (compensatory respiratory alkalosis).

KEY BOX 14.1

PRIMARY DISORDER	SECONDARY DISORDER
• Metabolic acidosis	• Respiratory alkalosis
• Respiratory acidosis	• Metabolic alkalosis
• Metabolic alkalosis	• Respiratory acidosis
• Respiratory alkalosis	• Metabolic acidosis

Thus, there are four primary disorders and four secondary disorders.

RESPIRATORY ACIDOSIS

Causes

This disorder occurs when the patient's ability to maintain minute ventilation is compromised. This may be acute or chronic in origin. The causes may be classified as follows:

- **Central nervous system:** Central nervous system depression due to trauma, tumour, infections, ischaemia or drug overdose. Spinal cord injuries, especially cervical or high thoracic can cause respiratory muscle paralysis.
- **Peripheral nervous and muscular system:** Guillain-Barré syndrome, tetanus, organophosphorus poisoning, poliomyelitis, myasthenia gravis.
- **Primary pulmonary disease:** Asthma, chronic obstructive pulmonary disease, acute respiratory distress syndrome, pneumonia.
- **Loss of mechanical integrity:** Flail chest.

Clinical features

- The features of the underlying problem predominate the clinical picture.
- If acute, hypoxia and hypercarbia result in tachycardia, hypertension, arrhythmias, confusion, drowsiness and coma. The hypoxia, if untreated, can be fatal.
- If gradual in onset, as in chronic obstructive pulmonary disease (COPD), the patient's kidneys may compensate by retaining bicarbonate resulting in compensatory metabolic alkalosis. Arterial blood gas analysis in these patients typically shows low PaO_2, high $PaCO_2$, high bicarbonate levels and a near-normal pH.

Treatment

- Treat the cause
- Maintenance of oxygenation and ventilation using mechanical ventilatory support till recovery of the primary problem occurs.

RESPIRATORY ALKALOSIS

This occurs due to an increase in minute ventilation. This increase can be sustained only in abnormal conditions. This may be acute or chronic in origin.

Causes (Key Box 14.2)

- Supratentorial lesions: Head injury
- Cirrhosis of liver
- Pain
- Anxiety, hysterical hyperventilation
- High altitudes
- It may also occur secondarily as a compensation to primary metabolic acidosis

KEY BOX 14.2

HYPERVENTILATION

- Head injury
- Hydrogen ions (metabolic acidosis)
- Hyperpyrexia
- Hysteria
- High altitudes

Features

- Usually features of the underlying disease predominate the picture.
- Acute severe hypocarbia ($PaCO_2$ < 20 mmHg) may cause cerebral vasoconstriction, reduced cerebral blood flow, confusion, seizures and tetany.
- The alkalosis and consequent hypokalaemia can also cause cardiac arrhythmias.

METABOLIC ACIDOSIS

Causes

This is associated with a decrease in bicarbonate ions due to one of two reasons:

1. ***Overproduction or retention of nonvolatile acids in the body,*** as in
   - Diabetic ketoacidosis
   - Lactic acidosis
   - Salicylate poisoning, methanol poisoning
   - Renal failure
2. ***Loss of bicarbonate ions from the body as in***
   - Diarrhoea
   - Intestinal fistulae

Features

- Usually features of the underlying disease predominate the picture.
- Hypotension, reduced cardiac output
- Hyperventilation—rapid, deep respirations
- Deep, gasping type of respiration seen in diabetic ketoacidosis is called **Kussmaul's respiration**.
- Hyperkalaemia, arrhythmias
- Lethargy, coma

Treatment

- Identify the cause and treat
- Adequate ventilation must always be ensured in all these critically ill patients.
- If pH < 7.1 and the patient is unstable, may administer sodium bicarbonate. The chances of life-threatening arrhythmias are reduced when pH is > 7.2.

Bicarbonate requirement (mmol) = Body weight (kg) × base deficit (mmol/L) × 0.3

(Each ml of 8.4% $NaHCO_3$ solution contains 1 mmol of HCO_3. Each ml of 7.5% $NaHCO_3$ solution contains 0.9 mmol of HCO_3^-).

Half the calculated dose of bicarbonate should be given slowly and should be followed up with repeat blood pH measurements as required.

ANION GAP

The law of electroneutrality states that the total number of positive charges must be equal to the total number of negative charges in the body fluids. Thus, cations (positively charged ions such as sodium and potassium) must produce a charge exactly balanced by anions. However, the concentrations of only sodium, potassium, chloride and bicarbonate ions are routinely measured in clinical practice. The sum of the measured cations (Na^+, K^+) exceeds the sum of measured anions (Cl^- and HCO_3^-) producing a 'deficit' called the "anion gap". The normal anion gap is 9–14 mmol/L. This gap is due to the presence of unmeasured anions in the body.

Since the extracellular concentrations of potassium is small, it is often ignored in the calculation of anion gap. The equation may be written as follows:

$$\text{Anion gap} = ([Na^+] + [K^+]) - ([Cl^-] + [HCO_3^-])$$

Anion gap may be used to distinguish the cause of metabolic acidosis.

- **Anion gap is increased (> 14 mmol/L) in metabolic acidosis due to an increase in fixed acid load.** These acids react with the bicarbonate ions in the plasma lowering its concentration. The anion portion of the fixed acid is not measured in the laboratory and contributes to the 'unmeasured anion' concentration, thus increasing the anion gap.
- **Anion gap remains unchanged in metabolic acidosis due to loss of bicarbonate ions** as the lost bicarbonate ions are replaced by chloride ions. This type of metabolic acidosis is also called "**hyperchloraemic acidosis**".

METABOLIC ALKALOSIS

Causes

This may be either due to loss of acid from the body or retention of bicarbonate. It may be due to:

- Loss of gastric hydrochloric acid as in vomiting, prolonged nasogastric drainage
- Excessive loss of H^+ from kidneys in exchange for K^+ in severe hypokalaemia
- Primary or secondary hyperaldosteronism
- Excessive exogenous administration of alkali, e.g. indiscriminate use of $NaHCO_3$, antacid abuse
- Retention of bicarbonate in exchange for loss of chloride ions as in diarrhoea.

Features

It is one of the common acid–base disorders in the intensive care unit. The underlying problem gives a clue to the cause of metabolic alkalosis. When severe, can cause hypoventilation and seizures. Associated hypokalaemia can cause arrhythmias and contribute to difficulty in weaning patients off a ventilator.

Treatment

- Treat the primary problem
- Most of the metabolic alkaloses are "chloride-responsive". Administration of saline and correction of potassium deficits reduce the alkalosis.
- Rarely, as in life-threatening metabolic alkalosis (pH > 7.7), rapid correction may be necessary and may be achieved by administration of H^+ in the form of dilute hydrochloric acid or ammonium chloride.

RAPID INTERPRETATION OF AN ABG REPORT

Analysis and conclusion of arterial blood gas (ABG) report must always be done in conjunction with history and clinical examination. ABG analysis is done to assess:

1. **Oxygenation status**
2. **Ventilatory status**
3. **Acid–base status**

Oxygenation

The partial pressure of oxygen in arterial blood (PaO_2) of a normal, healthy, young adult is usually 90–100 mmHg. An increase in inspired oxygen concentration (FIO_2) is expected to increase the PaO_2. The expected PaO_2 of a normal person may be estimated rapidly using the following formula:

$$\textbf{Expected } PaO_2 = FIO_2\ (\%) \times 5$$

For example: A person breathing 40% oxygen is expected to have a PaO_2 of $40 \times 5 = 200$ mmHg.

In patients with diseased lungs and increased shunt fraction (not ventilated but perfused alveoli—"wasted perfusion"), the PaO_2 will not rise at the same rate as in a normal person. As the shunt fraction increases, the rate of rise in PaO_2 reduces. If shunt fraction exceeds 40–50%, there may not be any rise in PaO_2 with an increase in FIO_2. Since the PaO_2 depends on the FIO_2, it is important to remember to relate the PaO_2 to the inspired oxygen concentration whenever oxygenation status of an individual is to be assessed.

Low PaO_2 responds to simple oxygen therapy if shunt fraction is 30% or less. If large shunts are present, measures must be taken to improve ventilation of the lungs, if necessary use endotracheal intubation and mechanical ventilation so that the shunt fraction is reduced.

A PaO_2 less than 60 mmHg can be life-threatening [corresponding to an arterial oxygen saturation, **SpO_2 of 90%**

as measured by a pulse oximeter and an attempt should always be made to keep a patient's PaO_2 (SpO_2) above this level]. Exceptions to this may be an individual acclimatised to low PaO_2 such as at high altitudes, cyanotic congenital heart disease or chronic obstructive pulmonary disease.

Nomograms are available to calculate the shunt fraction but an easy bedside assessment of oxygenation can be made using a PaO_2/FIO_2 ratio (Key Box 14.3). The ratio can be used to assess oxygenation and to evaluate the response to therapy. **A pulse oximeter is helpful** to assess whether the patient's oxygenation is life-threatening or not. A saturation of 98–100% may be reassuring. However, subtle changes in the oxygenation status may be missed if the patient is breathing high concentrations of oxygen. This is because of the sigmoid shape of the oxygen dissociation curve where the SaO_2 will be 99–100%, whether the PaO_2 is 100 mmHg or 500 mmHg. **Hence, an arterial blood gas analysis must always be obtained whenever a doubt exists about the oxygenation status of the patient.**

KEY BOX 14.3

PaO_2/FIO_2	STATUS OF OXYGENATION
500	Good
250–500	Acceptable
100–250	Poor
< 100	Critical

Ventilation

The **normal $PaCO_2$ is 35–45 mmHg**. The $PaCO_2$ must always be related to the alveolar ventilation of the patient. The minute volume of a normal healthy adult at rest would be 100 ml/kg/min. Sixty to sixty-five per cent of this actually ventilates the alveoli, the rest is dead space ventilation.

If the alveolar ventilation decreases (either due to a decrease in minute volume or an increase in dead space ventilation), the arterial $PaCO_2$ will rise. On the other hand, the arterial $PaCO_2$ may remain normal but the patient's minute volume may have increased.

Do not assume that the patient must be well when the $PaCO_2$ is normal. A clinical examination of the patient is necessary to rule out respiratory distress (dyspnoea, tachypnoea, active accessory muscles of respiration, tracheal tug, flaring of alae nasi, etc.). The patient may not be able to sustain this increased levels of ventilation for a prolonged period of time, is likely to get exhausted and may require mechanical ventilatory support.

Acid–base status

The assessment of acid–base status must be done in three steps and in the following order.

a. ***Assess the pH first:*** Normal pH—7.35 to 7.45. If the pH i less than 7.35, the patient has acidosis and if it is more thai 7.45, the patient is alkalotic. The direction of change in pH shows the primary disorder. This is because the compensatory mechanisms never overshoot the requiremen of reaching the normal pH.

b. ***Assess the $PaCO_2$ next:*** Is the $PaCO_2$ normal? The norma $PaCO_2$ is 35–45 mmHg. If the pH is abnormal but the $PaCO_2$ normal, it suggests a metabolic disorder. However, the body usually tries to compensate for a change in pH. The respiratory compensation is early and fast.

If the change in pH and $PaCO_2$ are in opposite directions (one is increased and the other decreased), the primary disorder is respiratory. If the change in pH and $PaCO_2$ are in the same direction (both are increased or decreased), the primary disorder must be metabolic.

For example, **if the pH is 7.2 and the $PaCO_2$ is 60 mmHg**, the decrease in pH suggests acidosis. The $PaCO_2$ has moved in the opposite direction (increased) and suggests respiratory acidosis. Since the direction of change of pH is towards acidosis and there is respiratory acidosis, it must be a primary respiratory disorder. Similarly, if the pH is alkalotic and the $PaCO_2$ is low, it suggests primary respiratory alkalosis.

The arterial carbon dioxide concentration ($PaCO_2$) and pH change in opposite directions. The extent and direction of change in pH due to a change in $PaCO_2$ depends on whether the disorder is acute or chronic and the amount of compensation. To analyse the disorder, the ratio of change in pH (ΔpH) to change in $PaCO_2$ ($\Delta PaCO_2$) can be used. ΔpH represents the difference between the normal pH (7.4) and the observed pH. $PaCO_2$ represents the difference between the normal $PaCO_2$ (40 mmHg) and observed $PaCO_2$.

See Key Box 14.4 for respiratory acidosis and respiratory alkalosis.

KEY BOX 14.4

RESPIRATORY ACIDOSIS

$\Delta pH/\Delta PaCO_2$	Condition
• > 0.008	Acute respiratory acidosis
• 0.008–0.003	Partially compensated respiratory acidosis
• < 0.003	Chronic (fully compensated) respiratory acidosis

RESPIRATORY ALKALOSIS

$\Delta pH/\Delta PaCO_2$	Condition
• > 0.008	Acute respiratory alkalosis
• 0.008 – 0.003	Partially compensated respiratory alkalosis
• < 0.003	Chronic (fully compensated) respiratory alkalosis

A change in $PaCO_2$ also changes the serum bicarbonate concentration. A rule of 1–4, 2–5 (given below) can be used to remember these changes (Key Box 14.5).

KEY BOX 14.5

Condition	PCO_2	$[HCO_3^-]$
• Acute respiratory acidosis	10 mmHg ↑	↑ 1 mmol/L
• Chronic respiratory acidosis	10 mmHg ↑	↑ 4 mmol/L
• Acute respiratory alkalosis	10 mmHg ↓	↓ 2 mmol/L
• Chronic respiratory alkalosis	10 mmHg ↓	↓ 5 mmol/L

c. Assess the bicarbonate level last: The normal plasma bicarbonate level is 22–26 mmol/L. An examination of the bicarbonate level after assessment of pH and $PaCO_2$ confirms the acid–base disorder. If the change in pH and the bicarbonate level is in the same direction, the disorder is primarily metabolic and vice versa. If the bicarbonate level is normal but the pH is acidotic, the disorder is of respiratory origin.

Changes in serum bicarbonate concentration is accompanied by compensatory changes in $PaCO_2$ (Key Box 14.6).

It must be remembered that these are general guidelines applicable to patients who are breathing spontaneously. These are useful for rapid bedside assessment of acid–base status. Occasionally, the patients can present with different combinations of acid–base disorders such as a mixed respiratory and metabolic alkalosis, or a mixed respiratory and metabolic acidosis. Such disorders are common in patients who are receiving mechanical ventilation in the intensive care unit.

The pH changes with a change in $PaCO_2$, the extent of which depends on whether the change is acute or chronic.

KEY BOX 14.6

Metabolic acidosis: Expected change in $PaCO_2$
$= \{\text{serum } [HCO_3^-] \times 1.5\} + 8 \pm 2$

Metabolic alkalosis: Expected change in $PaCO_2$
$= \{[HCO_3^-] \times 0.7]\} + 20 \pm 5$

CLINICAL NOTES

1. *A 30-year-old man was admitted to the ICU with history of consumption of organophosphorus poisoning 4 hours ago. On admission, he is drowsy, breathing 60% oxygen by face mask and has a bradycardia. A blood gas analysis taken half an hour later shows a PaO_2 = 100 mmHg, $PaCO_2$ = 60 mmHg and a pH of 7.24.*

Analysis

Oxygenation: *The PaO_2 on 60% oxygen should have been about 300 mmHg. The PaO_2/FIO_2 ratio in this patient is 100/0.6 = 167. Thus, although the PaO_2 is adequate to sustain life, the patient's oxygenation status is poor.*

Ventilation: *Raised $PaCO_2$ suggests respiratory acidosis.*

Acid–base status: *The pH shows acidosis. The pH has decreased, whereas the $PaCO_2$ has increased. Hence, the patient has primary, uncompensated respiratory acidosis.*

2. *A 60-year-old man, a known diabetic since the last 15 years is admitted with diabetic ketoacidosis. He required endotracheal intubation and ventilation with 60% oxygen. An arterial blood gas analysis shows the following:*

PaO_2 = 60 mmHg, $PaCO_2$ = 28 mmHg, pH = 7.14 and $[HCO_3^-]$ = 12 mmol/L

Analysis

Oxygenation: *The PaO_2 on 60% oxygen should have been about 300 mmHg. The PaO_2/FIO_2 ratio in this patient is 60/0.6 = 100. Thus, although the PaO_2 is just adequate to sustain life, the patient's oxygenation is poor.*

Ventilation: *Low $PaCO_2$ suggests respiratory alkalosis.*

Acid–base status: *The pH shows acidosis. The pH has decreased, whereas the $PaCO_2$ has also decreased. Hence, it is not respiratory acidosis and must be metabolic. The bicarbonate levels are far below normal and suggests a primary metabolic acidosis. The low $PaCO_2$ suggests secondary respiratory alkalosis. The patient has primary metabolic acidosis with partial compensation.*

3. *A 65-year-old man with a 40-year history of smoking, posted for elective herniorrhaphy was sent to the pre-anaesthetic clinic for evaluation. Since, he gave history of poor exercise tolerance as evidenced by breathlessness even on mild exertion, and clinical examination revealed presence of COPD, an arterial blood gas analysis was done while the patient breathed room air. The report showed a PaO_2 = 55 mmHg, $PaCO_2$ = 60 mmHg, pH = 7.34 and $[HCO_3^-]$ = 30 mmol/L.*

Analysis

Oxygenation: *The PaO_2/FIO_2 ratio is 55/0.21 = 262. Thus, although the PaO_2/FIO_2 ratio seems adequate, the actual PaO_2 is less than 60 mmHg and suggests hypoxaemia.*

Ventilation: *High $PaCO_2$ suggests respiratory acidosis.*

Acid–base status: *The pH shows acidosis. The pH has decreased, whereas the $PaCO_2$ is high. Hence, it is respiratory acidosis. The bicarbonate levels are high and suggests metabolic alkalosis. Since, the pH is acidotic but near normal, the patient must be having primary respiratory acidosis with compensatory metabolic alkalosis. He has fully compensated respiratory acidosis. This picture of chronic hypoxaemia and hypercarbia is typical of patients suffering from severe chronic obstructive pulmonary disease.*

When the serum bicarbonate levels change, there is a change in the arterial carbon dioxide levels also.

Whether a process is respiratory, metabolic or both, whether acute or chronic may be deduced from the history and physical examination of the patient. **Various nomograms** and formulae are available to evaluate these situations.

FLUIDS, ELECTROLYTES AND NUTRITION

A good understanding of the physiology of fluids and electrolytes is fundamental to the practice of surgery. Most surgical conditions are associated with changes in this balance and it is only appropriate that these are identified and treated effectively.

NORMAL PHYSIOLOGY

The human body consists of about 50–70% liquids and 30–50% solids by weight. The liquid portion varies with age, sex and body habitus. The variation is the result of individual differences in the fat content of the body which contains very little water. Hence, thin individuals have greater total body water (TBW) content as compared to obese individuals. Similarly, the TBW is about 50% in women and 60% in men. Neonates have up to 80% TBW. Of this total body water, intracellular water constitutes 40% of body weight (2/3 of TBW) and the extracellular portion, 20% of body weight (1/3 of TBW). The interstitial fluid and plasma portions of extracellular fluid constitute 15% and 5% of body weight respectively (Fig. 14.1).

Composition of body fluids

These fluid compartments are separated by semipermeable membranes allowing their fluid composition to be maintained within distinct limits. Table 14.1 shows the composition of the intracellular and extracellular fluid compartments. The composition of the intracellular compartments may vary according to the tissue, e.g. fat contains very little water.

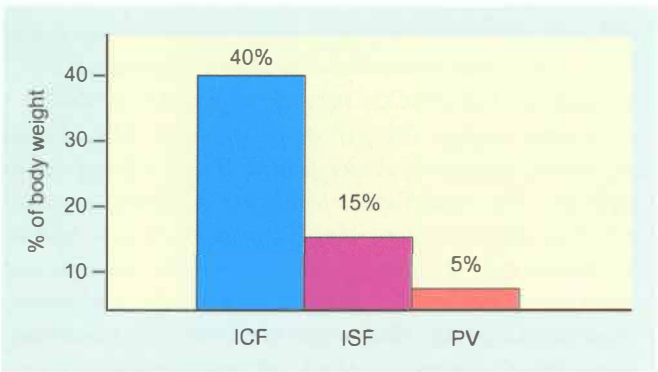


Fig. 14.1: Relation of various fluid compartments to body weight. ICF—intracellular fluid; ISF—interstitial fluid; PV—plasma volume

Table 14.1

Composition	Intracellular (mmol/L)	Extracellular (mmol/L)
Cations		
Sodium	10	140
Potassium	150	4
Calcium	2	2.5
Magnesium	20	1.5
Anions		
Chlorides	10	111
Bicarbonate	10	27
Sulphate	70	1.5
Phosphates	45	1

The tonicity of plasma is determined by the solutes, sodium and its corresponding anions, chlorides and bicarbonate, together with substances such as glucose, urea and proteins. These particles are osmotically active and hence, tonicity is described in terms of osmolality (mOsm/kg H_2O).

Osmolarity is concentration of a solution in terms of osmoles (or mosmoles) of **solute per litre of solution** (solute + water). **Osmolality** is concentration of a solution in terms of **osmoles (or mosmoles) per kilogram of solvent**. Osmolality is independent of the temperature of the solution and volume of the solute. Hence, osmolality is the preferred term in clinical practice.

Osmolarity: Osmoles per litre
Osmolality: Osmoles per kilogram

Osmolality of a solution can be measured in two ways:

1. ***By using the depression of freezing point of the solution:*** A solution of 1 Osm/kg freezes at –1.86°C. Normal plasma freezes at –0.54°C.

$$\textit{Plasma osmolality} = \frac{-0.54}{-1.86} \times 10^3 \text{ mOsm/kg}$$
$$= 290 \text{ mOsm/kg}$$

2. ***By estimating the solute concentration:*** Osmolality can be estimated from the concentration of major solutes of plasma.

$$\text{Osmolality} = 2 \times [Na^+] \text{ mmol/L} + \frac{[\text{Glucose (mg\%)}]}{18} + \frac{[\text{Blood urea (mg\%)}]}{6}$$

Example: If a patient's sodium concentration is 140 mmol/L, blood glucose concentration is 180 mg% and blood urea is 30 mg%, his plasma osmolality can be calculated as follows:

$$\text{Osmolality} = 2 \times [140] + \frac{180}{18} + \frac{30}{6}$$
$$= 280 + 10 + 5 = 295 \text{ mOsm/kg}$$

From the equation, it is evident that sodium contributes the most to the osmolality of plasma.

A change in osmolality is usually due to changes in sodium. The normal range of plasma osmolality is 285–300 mOsm/kg.

Plasma colloidal osmotic pressure

The plasma proteins normally do not pass out of the capillaries into the interstitium. These raise the plasma osmotic pressure above that of the interstitial fluid by an amount referred to as colloidal osmotic pressure (plasma oncotic pressure). The normal plasma colloidal osmotic pressure is 25 mmHg. Albumin is responsible for 75% of this oncotic pressure.

The body has mechanisms to regulate and maintain the volume of fluids, their concentration and composition within narrow limits to maintain homeostasis. Hence, a systematic assessment of fluid status of a patient involves the assessment of body fluid volume, its concentration and its composition in that order.

WATER REGULATION
(Regulation of Volume)

The primary methods of body water regulation are:

1. ***Regulating the volume of liquid ingested:*** When the extracellular fluid volume reduces, the thirst centre in the hypothalamus is stimulated which encourages the person to ingest more water.
2. ***Regulating the volume of urine excreted:*** This is regulated by plasma antidiuretic hormone (ADH). A reduction in plasma volume releases ADH from the posterior pituitary which in turn acts on the ADH receptors in the collecting tubules of the kidney. This results in increased reabsorption of water and reduced production of urine. ADH release may also be stimulated by increased plasma osmolality and angiotensin.

DISTURBANCES OF VOLUME

A decrease in the circulating volume is called hypovolaemia and an increase, hypervolaemia.

HYPOVOLAEMIA

This is common in surgical patients. The assessment of acute loss of blood volume is detailed in Chapter 12. The reduction in blood volume due to loss of water can be in the following ways:

a. Gut—vomiting, diarrhoea, fistulae
b. Skin and lungs—0.5 ml/kg/h normally, increases by 12% for every 1°C rise in temperature
c. Sequestration of fluid in third space.

Assessment of dehydration

This is a clinical assessment based upon

1. **History:** Severity and duration of loss of fluid.
2. **Examination:** Thirst, dryness of mucosa, loss of skin turgor, orthostatic hypotension, tachycardia, reduced jugular venous pressures and decreased urine output in the presence of normal renal function. Dehydration can be classified as given in Table 14.2.

Table 14.2

Degree of dehydration	Loss of body weight (%)	Clinical features
Mild	5	Reduced skin turgor, sunken eyes, dry mucous membranes
Moderate	10	Oliguria, hypotension and tachycardia in addition to the above
Severe	15	Profound oliguria and compromised cardiovascular function

Laboratory assessment

Haemoconcentration leads to falsely elevated haemoglobin, packed cell volume estimations and increased blood urea concentration. The kidneys reabsorb more water than usual leading to increased urine osmolality (> 650 mOsm/kg).

HYPERVOLAEMIA

Causes

1. Excessive infusion of intravenous fluids
2. Retention of water in abnormal conditions such as cardiac, renal and hepatic failure
3. Absorption of irrigation fluid as during transurethral resection of prostate using distilled water.

Diagnosis

- History and physical examination can lead to the cause.
- Physical examination: Distended neck veins, pedal oedema, body weight gain
- Circulatory overload:
  - Hypertension, tachycardia, pulmonary oedema
  - Confusion, restlessness, convulsions and coma

The development of these signs depends on the rate and volume of fluid overload, renal function and cardiovascular reserve.

Management

1. Treat the cause
2. Restriction of water and salt
3. Diuretics (or dialysis, if necessary) to remove excess water.

REGULATION OF SODIUM CONCENTRATION

Water constitutes the major component of all body fluids but the composition varies with the fluid compartment. The most abundant cation of extracellular fluid is sodium and is the prime determinant of ECF volume. Ninety per cent of the ECF osmolality is due to sodium.

The human body has no known mechanism to regulate sodium intake. The body regulates sodium output by:

- Regulating glomerular filtration rate
- Regulating plasma aldosterone levels

Addition or loss of water produces a change in the concentration of the solute. The quantity of solute relative to the volume of water is thereby increased (ECF is concentrated) or decreased (ECF is diluted) with loss or addition of water respectively. Changes in concentration are generally changes in water balance rather than changes in sodium regulation. Since the changes in volume and concentration are interdependent and the changes in water content are not easily measured, an estimate of the fluid volume and concentration is usually made by using the measured sodium levels and serum osmolality.

DISTURBANCES IN CONCENTRATION

HYPONATRAEMIA

Hyponatraemia is defined as a sodium level less than 135 mmol/L. It may occur as a result of water retention, sodium loss, or both. True hyponatraemia is always associated with low plasma osmolality. It may be associated with expanded, contracted or a normal extracellular volume.

Causes

Assessment of hypovolaemia should begin with an estimation of the extracellular fluid volume (clinically and if necessary, using central venous catheters). Thus, true hyponatraemia can be of three types: hypervolaemic hyponatraemia, hypovolaemic hyponatraemia and normovolaemic hyponatraemia (Fig. 14.2).

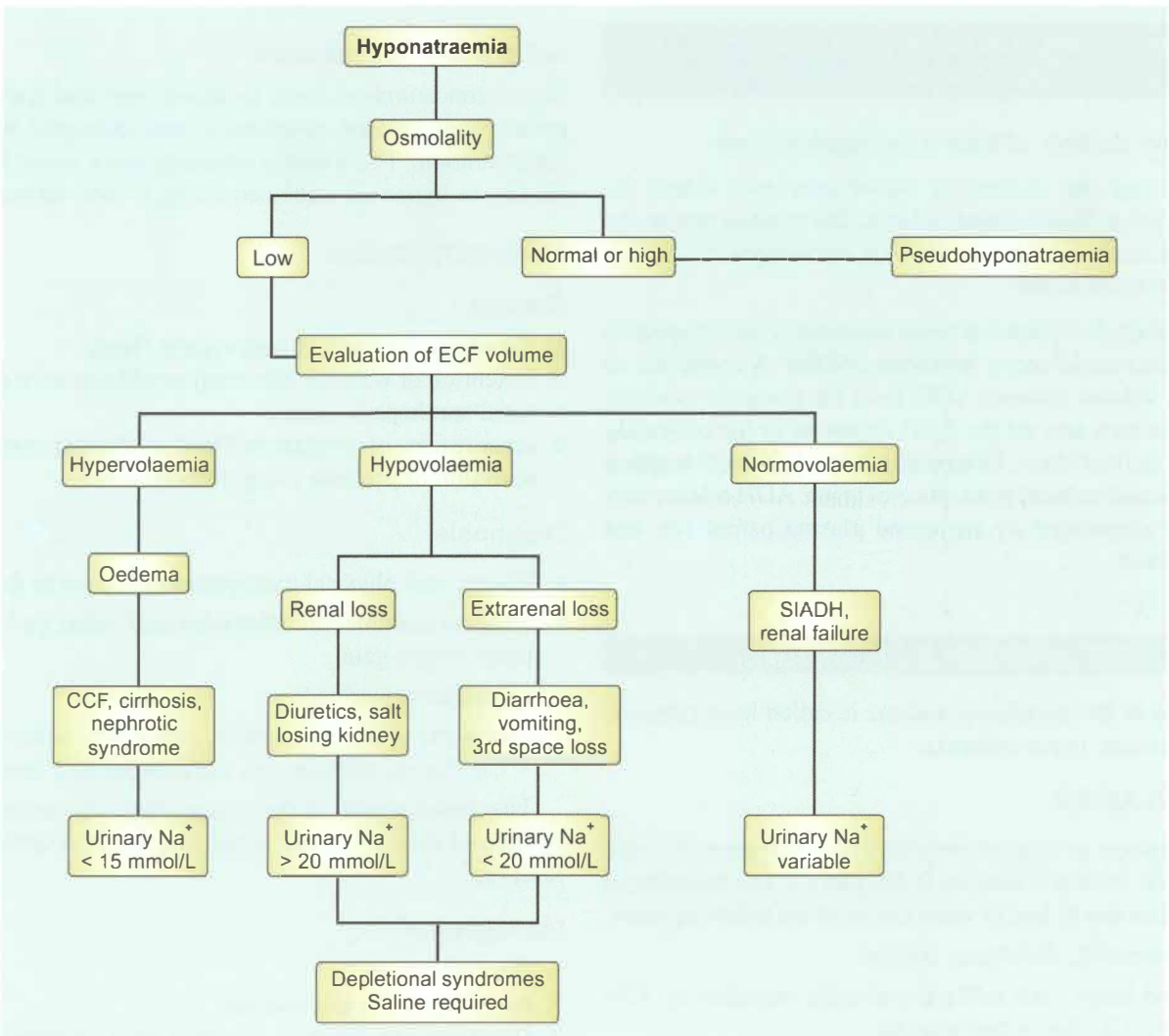


Fig. 14.2: Evaluation of hyponatraemia

I. Hypervolaemic hyponatraemia

Hypervolaemic hyponatraemia may be associated with clinical features of hypervolaemia such as oedema. Acute hypervolaemia (e.g. TURP syndrome—acute absorption of hypotonic fluids into the intravascular compartment) may result in cerebral oedema and pulmonary oedema. As plasma osmolality decreases, water moves from the extracellular space into the cells leading to oedema. The expansion of brain cells is responsible for the symptomatology of water intoxication: nausea, vomiting, lethargy, confusion, restlessness, etc. If severe ($[Na^+] < 100$ mmol/L), it can result in seizures and coma. Chronic hypervolaemia as in congestive cardiac failure, cirrhosis and nephrotic syndrome may manifest with pedal oedema and elevated jugular venous pressures till de-compensation occurs. The urinary sodium concentration is less than 15 mmol/L.

Treatment

Acute hyponatraemia (duration < 72 h) can be safely corrected more quickly than chronic hyponatraemia. The following factors must be evaluated: patient's volume status, duration and magnitude of the hyponatraemia and the degree and severity of clinical symptoms.

Fluid restriction, diuretics and correction of the underlying condition may be adequate in most cases. A combination of intravenous normal saline and diuresis with a loop diuretic (e.g. frusemide) also elevates serum sodium concentration.

Acute symptomatic hyponatraemia is a medical emergency. It should be treated with hypertonic saline (1.6% or 3%). Concomitant use of loop diuretics increases free water excretion and also decreases the risks of fluid overload.

The sodium concentration must be corrected to relieve symptoms and to a concentration of 125 mmol/L. Patients who are acutely symptomatic, the treatment goal is to increase the serum sodium by approximately 1–2 mEq/L/h until the neurologic symptoms subside. The correction should be slow and over a period of 12–24 hours with frequent checks of sodium concentration (every 2–4 h) to avoid overcorrection.

Avoid an absolute increase in serum sodium of more than 15–20 mEq/L in a 24-hour period. If sodium correction is undertaken too rapidly, the resulting osmolality changes in the extracellular fluid can cause central pontine myelinolysis. This condition is serious and can be irreversible.

The following equation can aid in the estimation of a sodium deficit to help determine the rate of saline infusion:

$$\text{Calculated sodium deficit} = (125 - \text{current serum } Na^+) \times (\text{body weight in kg}) \times 0.6$$

A litre of normal saline (0.9%) contains 154 mEq sodium chloride (NaCl) and 3% saline 500 mEq NaCl. In chronic severe hyponatraemia (i.e. serum sodium <115 mEq/L), the rate of correction should be slow and should not exceed 0.5–1.0 mEq/L/h, with a total increase not to exceed 10 mEq/L/day.

II. Hypovolaemic hyponatraemia

Hypovolaemia corrected inappropriately with hypotonic fluids such as 5% dextrose may result in hyponatraemia. The hypovolaemia may be due to renal causes such as diuresis or a salt-losing kidney. The urinary concentration of sodium is more than 20 mmol/L in these patients. Extrarenal loss of volume as in diarrhoea, vomiting or 3rd space loss may result in urinary concentration less than 20 mmol/L. All these are termed depletional syndromes and require saline infusion.

Treatment

Based upon the volume status, administer isotonic saline to patients with hypotonic hyponatremia who are hypovolaemic to re-expand the contracted intravascular volume.

III. Normovolaemic hyponatraemia

Occasionally, hyponatraemia may exist with normovolaemia. In such situations, the plasma osmolality must be estimated. If it is low, renal failure or the syndrome of inappropriate ADH secretion (SIADH) may be considered.

Treatment

For patients who have hypotonic hyponatraemia and are normovolaemic (euvolaemic), asymptomatic, and mildly hyponatraemic, water restriction (1 L/day) is generally the treatment of choice. For instance, a fluid restriction to 1 L/day is enough to raise the serum sodium in most patients. This approach is recommended for patients with asymptomatic SIADH.

Pharmacological agents can be used in some cases of more refractory SIADH, allowing more liberal fluid intake. Demeclocycline is the drug of choice to increase the diluting capacity of the kidneys by achieving vasopressin antagonism and a functional diabetes insipidus.

Pseudohyponatraemia

Occasionally, the hyponatraemia is only an apparent one due to the accumulation of other solutes such as glucose, urea, plasma proteins or lipids. The plasma osmolality is either high or normal in these patients. Such hyponatraemia is called pseudohyponatraemia.

Serum osmolality is governed by contributions from all molecules in the body that cannot easily move between the intracellular and extracellular space. Sodium is the most abundant electrolyte but glucose, urea, plasma proteins and lipids are also important. Normally, their concentrations are

small and contribute to the plasma osmolality only to a small extent. However, when the concentrations of these molecules increase to very high levels, the relative concentration of sodium in unit volume of serum may reduce. The actual amount of sodium is normal in these patients and hence the term pseudohyponatraemia. High blood sugar level or uraemia leads to higher plasma osmolality but high plasma protein or lipid levels is associated with normal plasma osmolality.

Treatment

The treatment of pseudohyponatraemia mainly involves treatment of the cause and supportive therapy.

HYPERNATRAEMIA

Hypernatraemia is defined as a plasma sodium concentration of more than 150 mmol/L and may result from pure water loss, hypotonic fluid loss or salt gain.

Causes of hypernatraemia

I. Pure water depletion

1. Extrarenal loss	Failure of water intake (coma, elderly, postoperative patients) Mucocutaneous loss—fever
2. Renal loss	Diabetes insipidus, chronic renal failure

II. Hypotonic fluid loss

1. Extrarenal loss	Gastrointestinal (vomiting, diarrhoea) Excessive sweating
2. Renal loss	Osmotic diuresis (glucose, urea, mannitol)
3. Salt gain	Iatrogenic (sodium bicarbonate, hypertonic saline), salt ingestion steroid excess

The hypertonicity of plasma leads to cellular dehydration. Clinical evidence of dehydration may not be apparent until 10–15% of body weight has been lost. Rehydration should be slow to prevent cerebral oedema.

The diagnosis can be established by measuring plasma and urine osmolalities and urine output.

- Uosm > Posm and ↓ urine output → Extrarenal causes (e.g. diarrhoea, fistulae.)
- Uosm > Posm and ↑ urine output → Osmotic diuresis
- Uosm < Posm and ↑ urine output → ↓ ADH or ↓ renal response to ADH.

Treatment

1. Administration of water orally/nasogastric tube
2. Administration of IV fluid—5% dextrose or 0.45% saline
3. Change in serum sodium should not be more than 2 mmol/L/h. Rapid rehydration can cause cerebral oedema.

DISTURBANCES IN COMPOSITION OF BODY FLUIDS

POTASSIUM BALANCE

The normal potassium level is 3.5–5.5 mmol/L. Hypokalaemia and hyperkalaemia are two clinically important disturbances.

Hypokalaemia (Key Box 14.7)

This is defined as a plasma concentration of potassium less than 3.5 mmol/L.

KEY BOX 14.7

CAUSES OF HYPOKALAEMIA

- Reduced intake
- Tissue redistribution: Insulin therapy, alkalaemia, β_2 adrenergic agonists, familial periodic paralysis.
- Increased loss: Gastrointestinal losses—diarrhoea, vomiting, fistulae
- Renal causes: Diuretics, renal artery stenosis, diuretic phase of renal failure

Symptoms

- Anorexia, nausea
- Muscle weakness, paralytic ileus
- Altered cardiac conduction: Delayed repolarisation, reduced height of 'T' wave, presence of 'U' wave, wide QRS complexes and arrhythmias.

Management

- Diagnosis and treatment of the cause
- Repletion of body stores
- Potassium supplements, in the form of milk, fruit juice, tender coconut water.
- Syrup potassium chloride orally—15 ml contains 20 mmol of potassium.
- If the patient cannot take orally or the hypokalaemia is severe, intravenous potassium chloride is usually given at a rate of 0.2 mmol/kg/h. If there are life-threatening arrhythmias, it may be given at a rate not exceeding 0.5 mmol/kg/h under electrocardiographic monitoring and serial measurements.

Hyperkalaemia

- This is defined as a plasma concentration of potassium more than 5.5 mmol/L.

Clinical features

- Vague muscle weakness, flaccid paralysis

Electrocardiographic changes

- Tall, peaked 'T' waves with shortened QT interval (6–7 mmol/L)

- Wide QRS complex, widening and then loss of 'P' wave (8–10 mmol/L)
- Wide QRS complex, merge into 'T' waves (sine wave pattern)
- Ventricular fibrillation (K^+ > 10 mmol/L)

Treatment of hyperkalaemia

1. Calcium gluconate (10%): 10–30 ml.
2. Sodium bicarbonate: 1–2 mmol/kg over 10–15 minutes.
3. 100 ml of 50% dextrose with 10–12 units of insulin over 15–20 minutes.
4. Hyperventilation
5. Salbutamol nebulisation
6. Calcium exchange resins
7. Peritoneal or haemodialysis

MAGNESIUM

- Magnesium is the second most abundant intracellular ion. The normal intracellular concentration of magnesium is about 26 mEq/L and extracellular concentration is 1.5–2.5 mEq/L. Almost 60% of the magnesium is deposited in the skeleton. Magnesium is required as a cofactor for several important enzymatic reactions including the phosphorylation of glucose within cells and the use of ATP by contracting muscle fibres.
- A daily dietary intake of 0.3–0.4 g (approximately 20–30 mEq) is required. The proximal convoluted tubule reabsorbs magnesium very effectively.

CALCIUM

- Calcium is the most abundant mineral in the body. Ninety-nine per cent is deposited in the skeleton. In addition, calcium ions are important for the control of muscular and neural activities, in blood clotting, as cofactors for enzymatic reactions and as second messengers.
- Calcium homeostasis reflects a balance between reserves in the bone, rate of absorption across the digestive tract, and rate of loss from the kidneys. The hormones parathyroid hormone (PTH), vitamin D and calcitonin maintain calcium homeostasis in the ECF. Parathyroid hormone and vitamin D raise Ca^{2+} concentrations and calcitonin lowers it.
- Calcium absorption from the digestive tract and reabsorption along the distal convoluted tubule are stimulated by PTH from the parathyroid glands and calcitriol from the kidneys. The average daily requirement of calcium in an adult is 0.8–1.2 g/day.

Hypercalcaemia (Key Box 14.8)

Hypercalcaemia exists when the Ca^{2+} concentration of the ECF is above 11 mg%.

Features

Severe hypercalcaemia (12–13 mg%) causes symptoms such as fatigue, confusion, cardiac arrhythmias, and calcification of the kidneys and soft tissues throughout the body.

Hypocalcaemia (Key Box 14.9)

Hypocalcaemia exists when calcium level is < 9 mg%. However, the serum calcium level should be related to the albumin levels. Half the serum calcium is bound to albumin and as albumin levels become low, this bound fraction is lower leading to a low total serum calcium concentration. Free ionic calcium is important for the electrical activity of the nerves and muscles and is more reliable (Normal: 1.0–1.4 mmol/L).

Features

Muscle spasms, stridor, generalised convulsions, myocardial depression, cardiac arrhythmia and osteoporosis.

CLINICAL NOTES

A 21-year-old lady was found to be collapsed as she was feeding her 15-day old baby in the nephrology ward. On arrival, the cardiac arrest response team found her to have ventricular tachycardia without pulse. Cardiopulmonary resuscitation was given and she was shifted to the intensive care unit after return of spontaneous circulation. Investigations showed that her potassium level was 1.6 mmol/L. She had been admitted to the nephrology unit for postpartum acute renal failure. She had been dialysed three times following which she had gone into the diuretic phase of recovery from acute renal failure. She was putting out about 5 litres of urine per day in the last two days. Her hypokalaemia was corrected over 2–3 days. She recovered completely and could be discharged from the ward in 5 days time.

KEY BOX 14.8

CAUSES OF HYPERCALCAEMIA

- Hyperparathyroidism
- Malignant cancers of the breast, lung, kidney or bone marrow

KEY BOX 14.9

CAUSES OF HYPOCALCAEMIA

- Hypoparathyroidism
- Vitamin D deficiency
- Chronic renal failure

PERIOPERATIVE FLUID THERAPY

A patient undergoing surgery needs intravenous fluids to replace volume deficit acquired during starvation, normal maintenance for the duration of surgery and volume lost during surgery. Depending on the extent of dissection, fluid accumulates in these tissues in the form of oedema (third space losses). In addition, blood loss also needs to be replaced.

Perioperative fluid therapy in a patient whose body homeostasis is normal

The replacement is as follows:

1. Fluid requirement during starvation: Patients awaiting anaesthesia and surgery need to be kept fasting for a few hours prior to and after the surgery. Except in very minor surgery, fluid lost during this period needs to be replaced. This volume is calculated at the rate of 2 ml/kg/h for the number of hours of fasting and is then replaced over 2–3 h.
2. Maintenance requirement: The average daily requirement of water for an average-sized adult is 2000 ml. In general, a volume of 30–35 ml/kg/day meets the daily maintenance needs. This is calculated as 2 ml/kg/h of surgery.
3. Third space losses:
   - 4 ml/kg/h for surgery with minimal dissection, e.g. herniorrhaphy
   - 6 ml/kg/h for surgery associated with moderate dissection, e.g. gastrojejunostomy and vagotomy
   - 8 ml/kg/h for surgery associated with large amount of dissection, e.g. Whipple's procedure.
4. Blood loss is replaced by compatible blood transfusion (homologous or autologous), if the haematocrit falls below 25%. Blood loss is replaced with an equal amount of colloids or three times the volume with crystalloids if the haematocrit is > 25% in an otherwise healthy individual. Crystalloids are electrolyte solutions that distribute themselves throughout the body water and hence, a larger volume needs to be given.

A patient undergoing surgery should receive fluid deficit due to starvation + maintenance fluids + third space losses + replacement of blood loss (as detailed above). Adequacy of fluid replacement should be checked with haemodynamic stability and urine output in major procedures. When very large fluid shifts are expected (oncologic surgeries), and the patient has compromised cardiac status or has renal insufficiency, it may be necessary to monitor fluid status using central venous pressure monitoring.

Perioperative fluid therapy in patients with disturbed fluid balance

Derangements of fluid therapy can be classified as:

a. Disturbances of volume
b. Disturbances of concentration
c. Disturbances of composition.

In the evaluation of a patient with a suspected problem in fluid and electrolyte or acid–base balance, careful sequential analysis of the volume, concentration and composition (in that order) followed by appropriate therapy protects the patient from severe, perhaps fatal errors in management.

Types of intravenous fluids

These can be broadly divided into three groups: crystalloids, colloids and special purpose solutions.

Crystalloids: These are essentially solutions of electrolytes in water, e.g. Ringer lactate. Some also contain dextrose, e.g. dextrose saline, 5% dextrose and paediatric maintenance solutions. They vary in the content of different electrolytes.

Colloids: These are solutions of large molecules which tend to remain in the intravascular compartment, e.g. gelatine, hetastarch, pentastarch, dextran 40, dextran 70. They are all plasma expanders since the molecules tend to exert osmotic forces and draw fluid from the interstitial compartment into the intravascular space. The colloids vary in their magnitude of volume expansion and duration of action. Dextran 40 reduces viscosity of blood and maintains blood rheology better. Hence, it is used as continuous infusion after microvascular surgery. It also reduces platelet aggregation which along with reduced viscosity helps in maintaining blood supply to free flaps and vascular grafts.

Volume deficits and losses are usually replaced using Ringer lactate. However, if the patient has disturbances of concentration and composition, other solutions may need to be given.

SPECIAL PURPOSE SOLUTIONS

Sodium bicarbonate: It is available as 7.5% (0.9 mEq/ml) and 8.4% (1 mEq/ml) of sodium bicarbonate.

Uses

- As an alkalinising agent
- To treat metabolic acidosis
- To treat hyperkalaemia
- Forced alkaline diuresis.

Disadvantages

- Increased sodium load
- Alkalosis with a shift of oxygen dissociation curve to the left (increased affinity of haemoglobin to O_2, reducing its unloading)
- Increased intracranial pressure and intraventricular haemorrhages in neonates

- Circulatory overload leading to cardiac failure
- Carbon dioxide load leading to respiratory failure

Mannitol (10 and 20%): It is an osmotic diuretic. Its main use is to reduce intracranial pressure by producing diuresis. It is also used to reduce intraocular pressure. Mannitol expands intravascular volume initially by drawing fluid from the interstitium. This is followed by diuresis. It should be used with caution in patients with cardiac failure, renal failure, etc.

Hypertonic saline (1.6, 3, and 5%): These solutions are available to treat hyponatraemia.

Albumin: 5% albumin is used as a plasma expander. 20% albumin can be used to replace lost albumin in severe hypoalbuminaemia in addition to plasma expansion.

NUTRITION

Nutritional support to patients

Cells can perform their function only when they get nutrients and oxygen. When the nutrients are metabolised in the cells, the tissues get energy to perform their physiological functions. Gastrointestinal system (GIT) is the source of supply of nutrients (energy) to all the tissues. It has to supply nutrients on a day-to-day basis because the body has limited expendable reserves (stores). Lack of nutrients results in energy crisis.

In critical care units, priority is given to treatment of hypoxia, haemorrhage, haemodynamic instability, fluid, electrolyte, acid–base imbalance, and sepsis. These deserve their priority, but hyponutrition and the consequent energy crisis should not be ignored.

- **Pathophysiology of GI failure—autocannibalism:** When GIT fails, the stores are consumed to supply energy. Glycogen stored in the liver undergoes glycogenolysis and gets depleted in 24 to 48 hours. Normal physiological functions of the organs of an average adult at rest (BMR/Resting energy expenditure—REE) need about 20 kcal/kg/day. Therefore, when GIT fails, the fat in the adipose tissue and protein in the muscles and viscera are mobilised and metabolised to supply and sustain REE. This is called autocannibalism (eating one's own tissues to survive). It weakens the muscles (e.g. respiratory and cardiac muscles), viscera (liver, kidneys, etc.) and immune system, resulting in increased morbidity and mortality.
- **Patients receiving intravenous (IV) fluids are semistarving:** 500 ml of 5% dextrose containing 25 g of dextrose provides about 100 kcal (each gram of carbohydrate provides about 4 kcal), and therefore an adult taking nil by mouth (NBM), receiving 4–5 bottles of 5% dextrose/day gets about 400–500 kcal; the balance amount of REE (for a 50-kg individual, REE = 50 × 25 = 1250 kcal; 1250 – 500 = 750 kcal) comes from glycogen for 24 to 48 hours and thereafter from autocannibalism which is detrimental.
- **Fasting in healthy persons versus fasting in patients:** When resting healthy persons are fasting, the metabolic rate drops to basal level. Although critically ill patients who are kept NBM are also resting, their basal metabolic rate is accelerated (hypermetabolic) and their REE is increased proportionate to the level of stress due to injury or illness.
- **Assessment of nutritional status:** A healthy adult can withstand semistarvation (receiving IV fluids only) for about 4 to 5 days without ill-effects. Beyond that they need nutritional support in order to prevent the adverse effects of autocannibalism. Many patients are nutritionally depleted at the time of admission and will need nutritional support much earlier.
- Weight loss is an important indicator of nutritional status. A recent h/o 20% weight loss indicates mild, 20 to 40% moderate and more than 40% severe undernutrition. Body mass index (BMI) is another easily calculated indicator. Mid-arm circumference, triceps skin fold thickness are anthropometric indices that are useful. Low serum albumin (3–3.5 g%—mild, 2 g%—moderate and < 2 g%—severe undernutrition) is not a sensitive indicator because of the long half-life and a large pool size. Prealbumin, retinol-binding protein and transferrin have shorter half-life and smaller pool size and are more sensitive indicators.
- **How much nutrition to be given:** Resting energy expenditure (REE) can be more accurately calculated by the Harris-Benedict equation. The REE × stress factor equals total caloric requirement. The stress factor depends upon the severity of illness, injury or operation and ranges from 1.2 to 2.0. For practical purposes, it is easier to remember that the adult REE is 20 kcal/kg/day and it increases to 25, 30, and 40 kcal/kg/day in mild, moderate and severe stress.
- Sixty per cent of the total calories should come from carbohydrates and 40% from fats. These calories are nonprotein calories. Calories obtained from proteins should not be taken into account for calculating the energy needs because they are building blocks in tissue repair and are not meant for burning for calories. The protein requirement of the resting adult is 0.8 g/kg/day, and it increases to 1.1, 1.5, and 2.0 g/kg/day in mild, moderate and severe stress. Recommended daily allowances of vitamins, minerals and trace elements are added to the formulations. The daily intake and output of fluids should be balanced.
- Nutrition can be administered by either the enteral or parenteral route.

Enteral nutrition

It is paradoxical that sick patients who need to eat more to meet the increased metabolic demands are often unable to eat.

They have anorexia, nausea, vomiting and altered sensorium. Oral feeding is impossible in patients with faciomaxillary injuries or those on ventilators. In many of them, the intestines are working. Enteral route is best for providing nutrition. Hence the dictum, **"When the gut is working, use it".**

- **Enteral Access:** Following are the routes to introduce nutrients into the GIT (Table 14.3):
  - ***Nasogastric feeding:*** When the stomach emptying is normal and swallowing is impossible or contraindicated, nasogastric feeding (Ryle's tube) provides nutrition.
  - ***Nasojejunal feeding:*** In gastric stasis, feeding can be given through a nasojejunal (NJ) feeding tube introduced either blindly, or under radiologic or endoscopic guidance to place its tip in the jejunum (postpyloric).
  - ***Feeding gastrostomy:*** By open method or by percutaneous endoscopic gastrostomy (PEG) when RT or NJ tube insertion is impossible.
  - ***Percutaneous Endoscopic Gastrostomy (PEG):*** With the help of an endoscope, a gastrostomy tube is placed in a retrograde manner and brought out through a skin incision. It is technically very easy and can be done under local anaesthesia. It has replaced feeding gastrostomies (open method). It is popular nowadays (Fig. 14.3). Complications include colonic perforation, sepsis, bleeding, wound infection, etc.
  - ***Feeding jejunostomy:*** After major/complex operative procedures on the oesophagus, stomach and pancreas, a feeding jejunostomy is frequently established.
- **What to feed:** A number of preparations are commercially available but most cost-effective ones are the blenderised kitchen feeds. Enteral feeds are hyperosmolar and provide 1.2 to 2.0 kcal/ml.
  - ***Polymeric feeds:*** These are commonly prepared in the kitchen. Liquid and powder preparations are commercially produced. These contain polysaccharides polypeptides and oils. Soups of dal, vegetables and chicken are examples of polymeric feeds.
  - ***Elemental feeds:*** These are predigested *in vitro* and contain oligosaccharides, oligopeptides and medium and long chain triglycerides (MCT and LCT). They are useful in patients with irritable bowel disease and short bowel.
  - ***Modular feeds:*** Contain monosaccharides, amino acids and fatty acids.
  - ***Disease-specific feeds:*** The composition of the feeds needs to be altered in certain disease states. Renal failure—low protein, low/ no electrolytes; Hepatic failure—more branched chain amino acids (BCAA) and less aromatic amino acids; Respiratory failure—more fats (55% cal) and less carbohydrates.
- **How to feed:** The feeds can be gravitated, injected with a syringe or pumped into the tubes either continuously or intermittently. To start with 50 ml every two hours on the first day and if tolerated may be increased gradually to 200 ml every two hours until the target is reached.

Table 14.3 Feeding methods (Figs 14.4 and 14.5)

Ryle's tube (RT) feeding	Gastrostomy	Feeding jejunostomy
Easy, quick, cheap method Indicated in stroke, comatose patients, etc.	Indicated when RT cannot be passed, e.g. inoperable carcinoma oesophagus, stricture	Indicated after major oesophageal surgeries, high duodenal fistulae
Chances of aspiration are high. Hence, 30° propped up position is recommended	Malecot's catheter is introduced into the stomach and kept in place using a purse string suture (Stamm's gastrostomy)	A Ryle's tube is introduced into the jejunum under vision (during surgery) and kept in place using a purse string suture

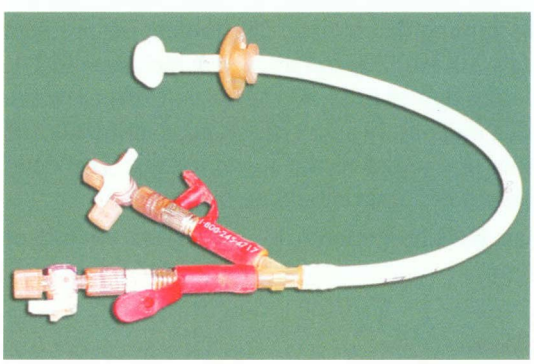


Fig. 14.3: Percutaneous endoscopic gastrostomy tube

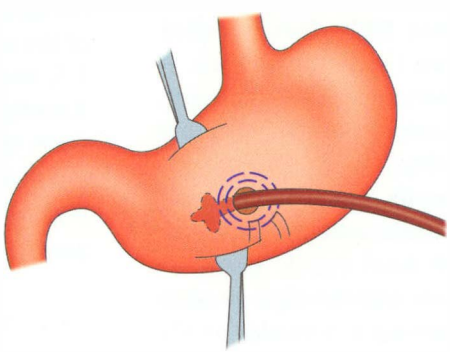


Fig. 14.4: Feeding gastrostomy

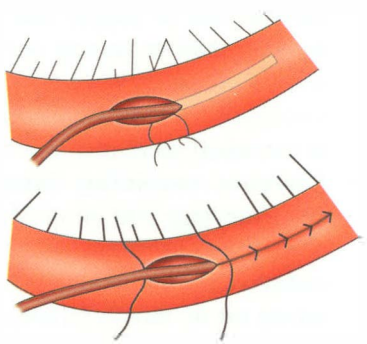


Fig. 14.5: Feeding jejunostomy

- **Advantages of enteral nutrition:**
  1. **The integrity of gut mucosa depends on provision of nutrients into the gut lumen.** If the fasting period exceeds more than a few hours, the gut mucosal cells start disintegrating and the villi get destroyed. This may permit the intestinal bacteria to enter the circulation leading to sepsis. Translocation of bacteria from the intestines into the circulation has been identified as the **'motor of multiorgan failure'**.
  2. Use of natural route of nutrition requires less nursing supervision.
  3. Infection rate is lower with enteral nutrition.
  4. Greater insulin response is seen with enteral nutrition.
  5. There is a lower tendency to retain salt and water.
  6. Enteral nutrition is cheaper.
- **Complications of enteral nutrition:** Nausea, vomiting, abdominal distension and diarrhoea are common. However, intractable diarrhoea should be investigated for *Clostridium difficile* infection. Electrolyte imbalance, hyperosmolar coma, refeeding syndrome and aspiration are other problems. Tube clogging, displacement, leak and erosion are mechanical complications.

Immunonutrition

Glutamine is an essential nutrient for the maintenance of integrity of intestinal cells and there is evidence to show that its inclusion in enteral feeds enhances patient immunity.

PARENTERAL NUTRITION

When enteral nutrition is not possible for more than a few days, parenteral nutrition (PN) may need to be considered. When all nutrition is done by the parenteral route, it is termed total parenteral nutrition. Partial parenteral nutrition may be given to supplement inadequate enteral nutrition.

Parenteral nutrition (PN) to GI failure is like dialysis to renal failure and ventilator support to respiratory failure. When enteral feeding is impossible, parenteral nutrition should be given. Prolonged ileus, intestinal obstruction, malabsorption, short gut, inflammatory bowel disease, high output intestinal fistulae are some common indications for PN.

PN formulations are marketed as

1. Dextrose (10, 20, 25 and 50%), amino acid (5 and 10%) and lipid (10 and 20%) solutions in separate bottles.
2. Dextrose + amino acid solutions
3. All in one solutions.

Insulin is commonly added to PN solutions. Heparin is occasionally added.

Administration of nutrients

Carbohydrates

One gram of glucose provides 4 kcal. The entire energy requirement of the day can be given in the form of glucose. This needs to be administered in a concentration of 50%, so as not to exceed the daily fluid requirement. 1000 ml of 50% dextrose contains 500 g of dextrose providing 2000 kcal.

Problems

- Hyperglycaemia must be prevented by addition of appropriate amount of insulin.
- Potassium supplements need to be added to avoid hypokalaemia.
- Increased CO_2 production may increase work of breathing and results in difficulty in weaning patients from ventilator.
- Hypertonic solutions produce thrombophlebitis
- Infusion of dextrose-containing solutions do not take care of daily protein and other nutrient requirements of the body.

Thus, parenteral nutrition should not be given using dextrose alone.

Lipids

One gram of fat provides 9 kcal. Since, it is a more concentrated form of energy, half the daily caloric requirement can be given using 10 or 20% lipid emulsions.

Problems

- Occasionally lipid infusion can cause impaired pulmonary diffusing capacity.
- Its clearance from plasma may be delayed with impaired liver function.
- Lipid emulsions are expensive.

Proteins

The average protein requirement is 1 g/kg/day. Amino acid replacement solutions are available as 10 and 20% solutions. The calorific value of proteins should not be counted as they are the building blocks of the body. They are not infused to be metabolised for energy.

Electrolyte requirements

The daily requirement of various electrolytes are given below.

Electrolyte	mmol/kg/day
Sodium	1–2
Potassium	1
Calcium	5–10
Magnesium	5–10

Vitamins are given separately. Trace elements are given weekly to patients on long-term PN. One ampule of water-soluble vitamins must be infused daily, over a period of time exceeding 30 minutes to avoid urinary loss. Folic acid, vitamins B_{12}, K, A and D also need to be given once a week.

The total volume of PN should be 2000–3000 ml/day. Alternately, commercially available PN solutions which are well-balanced with all nutrients can be used. Although expensive, good nutrition may reduce the morbidity and duration of hospital stay of the patient thus proving cost-effective.

Routes of administration

1. **Peripheral vein:** Solutions with less than 800 mOsm/L may be administered through a peripheral vein. This is suitable for short-term PN.
2. **Central vein:** Either internal jugular or subclavian vein is cannulated. PN solutions with higher osmolality must be given through a central vein.
3. **Peripherally inserted central vein catheter:** This also may be used for short-term purposes.
4. **Subcutaneously implanted central vein catheter** ports are especially suitable for long-term, domestic or ambulatory PN.

PEARLS OF WISDOM

The central venous access should be dedicated to PN and should not be used for administration of drugs or other fluids. The line should be handled with strict asepsis.

Methods of administration

- The solutions can be gravitated but the rate of infusion is better controlled if given through pumps. Smaller volumes are given initially and is gradually increased to reach the target volume/day. Absolute aseptic precautions are observed while handling the catheters and the PN formulations, since central vein catheter infection is a dangerous complication. Central vein catheter should be used exclusively for administering PN solutions and should not be used for any other purpose.

Monitoring

Aim

1. To identify excess or deficiency of individual nutrients.
2. To identify complications.

Daily

- Blood sugar, serum electrolytes, blood urea and serum creatinine.

Biweekly

- Liver function tests, coagulation profile, complete haemogram.

Complications of PN

1. **Technical complications:** Injury to subclavian/carotid artery, brachial plexus, haemo or pneumothorax.
2. **Catheter-related:** Central line sepsis is the most dangerous, at times, life-threatening and yet, preventable complication and its incidence is a measure of patient safety. It may not be possible to control sepsis with antibiotics alone without removing the central catheter. Thrombosis and catheter clogging are other problems.
3. **Gut mucosal atrophy:** Patients on total parenteral nutrition develop atrophy of the intestinal mucosa which loses its barrier function and becomes permeable to bacteria. The consequent bacterial translocation leads to sepsis and multiorgan dysfunction syndrome (MODS). This is not seen in patients on partial PN supplemented with EN.
4. **Cholestasis:** Some patients on long-term TPN develop cholestasis, jaundice and gallstones which resolve on starting oral/EN.
5. **Fluid, electrolyte and acid–base imbalances** are common.

Assured delivery of nutrients, accurate and rapid correction of fluid, electrolyte and acid–base imbalances are the merits of PN but it is complicated and expensive.

PEARLS OF WISDOM

Nutrition is an important component of the care of the surgical or critically ill patient. Malnutrition can be fatal. Overnutrition can also be harmful. Provision of services of a physician, dietician, microbiologist and good nursing care are vital to patient recovery.

MULTIPLE CHOICE QUESTIONS

1. The normal hydrogen ion concentration of plasma is _____ nmol/L:

A. 30 B. 40
C. 50 D. 60

2. The most important buffer system in the plasma is:

A. Phosphate buffer system
B. Ammonia buffer system
C. Proteins
D. Bicarbonate-carbonic acid buffer system

3. pH is the negative logarithm to the base 10 of hydrogen ion concentration expressed in _____.

A. mol/l B. mmol/L
C. nmol/L D. µmol/L

4. The solubility coefficient of carbon dioxide in plasma is _____ ml/mm Hg/dL

A. 0.3 B. 0.03
C. 0.003 D. 3

5. The following is true in primary metabolic acidosis with secondary respiratory alkalosis:

A. pH and PCO_2 are decreased
B. pH decreases but $PaCO_2$ increases
C. pH and $PaCO_2$ are increased
D. pH increases but $PaCO_2$ decreases

6. The following is one of the causes of metabolic acidosis with increased anion gap:

A. Diarrhoea B. Intestinal fistula
C. Pancreatic fistula D. Diabetic ketoacidosis

7. The following equation represents the nonlogarithmic form of acid–base equation:

A. Harrison
B. Henderson
C. Hutchinson
D. Hanson

8. The following may be a cause of respiratory alkalosis:

A. Morphine overdose
B. Chronic obstructive pulmonary disease
C. Salicylate poisoning
D. Curare poisoning

9. In the normal oxyhaemoglobin dissociation curve, a saturation of 90% corresponds to _____ mm Hg PO_2.

A. 40 B. 60
C. 80 D. 120

10. Metabolic acidosis with normal anion gap is also called:

A. Hyperchloraemic acidosis
B. Hyperkalaemic acidosis
C. Hypernatraemic acidosis
D. Hypercalcaemic acidosis

11. The following electrolyte contributes most to the osmolality of plasma:

A. Sodium
B. Potassium
C. Magnesium
D. Calcium

12. The normal plasma osmolality is _____ mOsm/kg:

A. 190 B. 290
C. 160 D. 260

13. Plasma osmolality is determined in the laboratory using:

A. Freezing point
B. Boiling point
C. Saturation point
D. Isoelectric point

14. The second major intracellular cation is:

A. Sodium
B. Potassium
C. Calcium
D. Magnesium

15. In chronic hyponatraemia, the sodium concentration should be increased at a rate not exceeding _____ mmol/L/h.

A. 1
B. 5
C. 10
D. 15

16. The following is one of the drugs used to treat hyperkalaemia:

A. Digoxin
B. Magnesium sulphate
C. Atropine
D. Insulin

17. Tall 'T' wave in the electrocardiogram is a feature of:

A. Hypokalaemia
B. Hyperkalaemia
C. Hypocalcaemia
D. Hypercalcaemia

18. Rapid infusion of the following fluid can cause intraventricular haemorrhage in neonates:

A. 5% dextrose
B. 8.4% sodium bicarbonate
C. 0.9% saline
D. Ringer lactate

19. Translocation of bacteria from the following organ can lead to multiorgan failure and thus this organ is called 'motor of multiorgan failure':

A. Kidney
B. Brain
C. Gut
D. Liver

20. Hypercalcaemia may be seen in the followin conditions *except:*

A. Hyperparathyroidism
B. Malignant cancers of the breast and lung
C. Vitamin D toxicity
D. Chronic renal failure

ANSWERS

1 B	2 D	3 A	4 B	5 A	6 D	7 B	8 C	9 B	10 A
11 A	12 B	13 A	14 D	15 A	16 D	17 B	18 B	19 C	20 D

15

Tumours and Soft Tissue Sarcoma

- Benign tumours
  - Papilloma
  - Fibroma
  - Lipoma
- Neural tumours
  - Neuroma
  - Neurofibroma
  - Neurilemmoma
  - Chordoma
- Malignant tumours
  - Paraneoplastic syndromes
- Soft tissue sarcomas
  - Differential diagnosis of soft tissue sarcoma
  - Liposarcoma
  - Malignant fibrous histiocytoma
  - Synovial sarcoma
  - Angiosarcoma
  - Rhabdomyosarcoma
  - Kaposi's sarcoma
  - Dermatofibrosarcoma protuberans

Introduction

A tumour is a new growth consisting of cells of independent growth arranged atypically and serves no function. Broadly classified into:

- Benign
- Malignant

BENIGN TUMOURS

PAPILLOMA

This is a benign tumour arising from skin or mucous membrane. It is characterised by finger-like projections with a central core of connective tissue, blood vessels, lymphatics and lining epithelium (Fig. 15.1). It can be called Hamartoma or a skin tag. It is an example of overgrowth of fibrous tissue (Key Box 15.1). It can be pedunculated with narrow base or broad base.

PEARLS OF WISDOM

Acrochordons (skin tags) are fleshy, pedunculated masses located on the axillae, trunk and eyelids.

KEY BOX 15.1

OVERGROWTH OF FIBROUS TISSUE

- Keloid : Page 8
- Desmoid tumour : Page 910
- Hypertrophic scar : Page 8

Types

1. Skin papilloma

a. ***Squamous papilloma*** occurs in the skin, cheek, tongue, etc.

- **Soft papillomas** are squamous papillomas. They are seen in elderly patients on the eyelid as small, soft, brownish swellings.
- **Squamous papilloma** can also be ***congenital,*** sometimes multiple in number and can be sessile or pedunculated.

b. ***Basal cell papilloma*** (seborrhoeic keratosis) is seen on the trunk of elderly patients as brownish elevated patch of skin and gives a semitransparent, oily appearance.

2. Arising from mucous membrane of visceral organs

a. ***Transitional cell papilloma*** in the urinary bladder as a cause of haematuria.

b. ***Columnar cell papilloma*** in the rectum as a cause of mucous diarrhoea.

c. ***Cuboidal cell papilloma*** in the *gall bladder.*

d. ***Squamous papilloma*** in the larynx can cause *respiratory obstruction.*

e. ***Papilloma of breast*** **(duct papilloma) causes bleeding per nipple.**

Treatment

- Excision, only if papilloma causes discomfort, or if it is symptomatic.

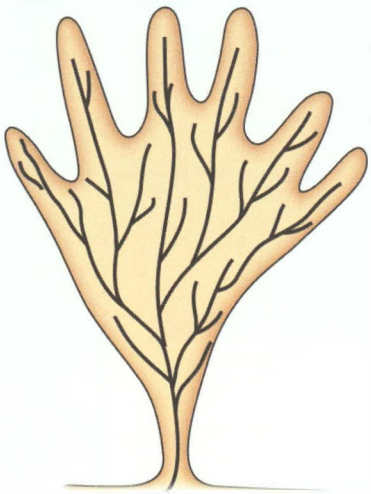


Fig. 15.1: Structure of papilloma

Fig. 15.2: Pedunculated papilloma thigh—broad base

Fig. 15.3: Papilloma thigh with a narrow base—easy to remove it. Ulceration can be a problem here

Complications

1. Skin papilloma can get secondarily infected resulting in pain and swelling.
2. Ulceration and bleeding (trauma)
3. Papilloma in the breast, rectum, tongue and gall bladder can undergo malignant change.

FIBROMA[1]

Fibroma is a benign tumour, consisting of connective tissue fibres only. Clinically, it presents as a firm, subcutaneous swelling. However, ***a true fibroma is rare.*** They are combined with neural elements, muscle tissue or fatty tissue.

Types

1. Soft fibromas: Less fibrous tissue
2. Hard fibromas: More fibrous tissue
   - Neurofibroma: Fibroma mixed with nerve fibres
   - Fibrolipoma: Fibroma mixed with fat
   - Myofibroma: Fibroma mixed with muscle fibres
   - Angiofibroma: Fibroma mixed with blood vessels

Treatment

They are treated by excision because of the possibility of developing into a sarcoma.

LIPOMA: UNIVERSAL TUMOUR

Lipoma is a benign tumour arising from fat cells of adult type. It is also called 'universal tumour' because it can occur anywhere in the body where there is fat (Key Box 15.2).

Types

1. Single encapsulated lipoma

- This is a single, soft, slow-growing, painless and semi-fluctuant swelling (Figs 15.4 and 15.5).

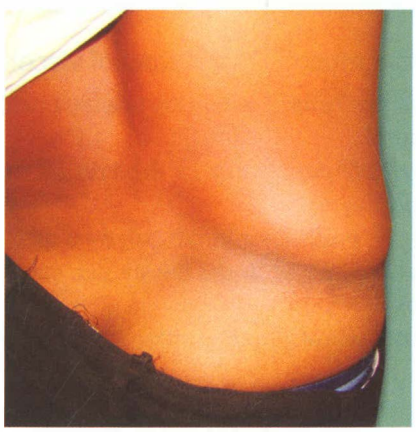


Fig. 15.4: Lipoma in the flank—**commonest site of lipoma**

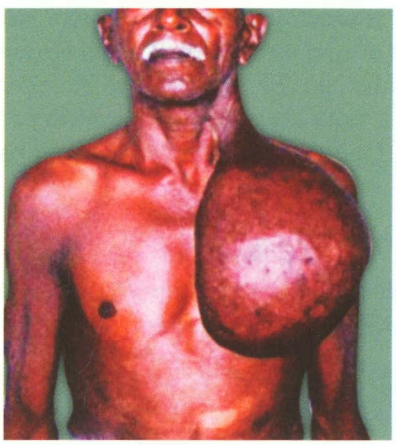


Fig. 15.5: Giant lipoma in the chest wall (*Courtesy:* Prof Rajiv Shetty, HOD, Bangalore Medical College, Medical Superintendent, Bowring Hospital, Bangalore)

[1]Students should not give the diagnosis of fibroma because in majority of cases, it is neurofibroma or fibrolipoma.

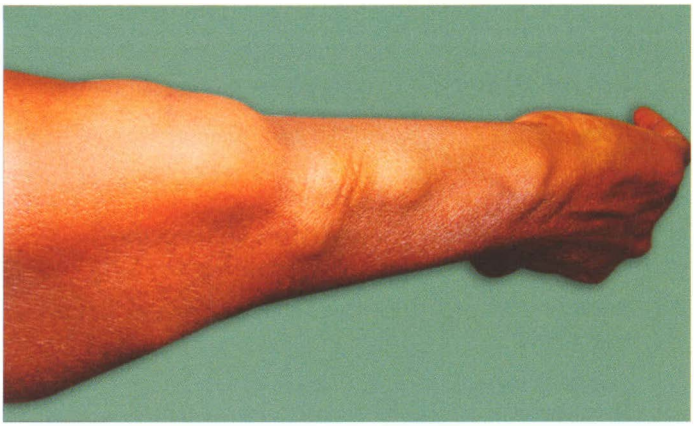


Fig. 15.6: Multiple lipomas—being subcutaneous in location, they become **prominent on contraction of muscles**

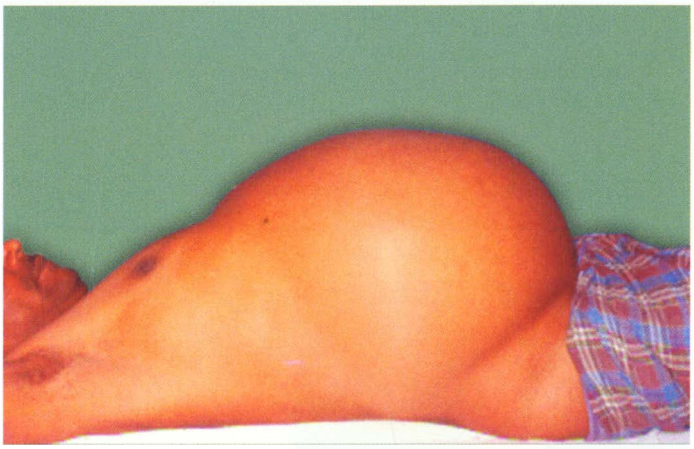


Fig. 15.7: Retroperitoneal lipoma. It can grow to **large dimensions.** Mostly asymptomatic, and hence patients come late to the hospital

KEY BOX 15.2

DIAGNOSTIC FEATURES OF LIPOMA

- Subcutaneous—commonest type
- Soft to firm lobular swelling
- 'Slip' sign positive—a pathognomonic sign
- Semifluctuant swelling
- 'Smart' dimple sign on movement of the skin

- The swelling is soft, may feel cystic with fluctuation. This is also called pseudofluctuation because fat at body temperature behaves like fluid.
- Surface is **lobular.** Lobulations are better appreciated with firm palpation of the swelling. Due to the pressure, lobules bulge out between the fibrous tissue strands.
- ***The edge slips under the palpating finger which is a pathognomonic sign of lipoma.***

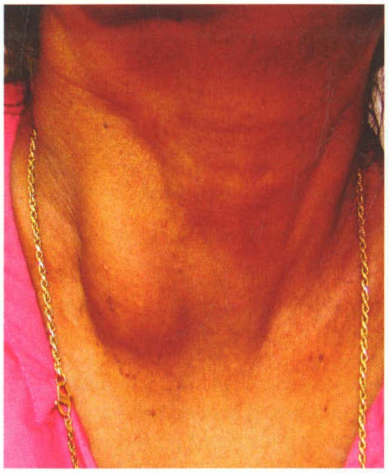


Fig. 15.8: Mediastinal lipoma. The swelling was deep to pretracheal fascia and partly substernal. It was soft and lobular—confused for compressible swelling. Candidate offered haemangioma as diagnosis. (*Courtesy:* Dr Salim, HOD, Surgery, Trivandrum Medical College, Kerala (PG exam case 2009)

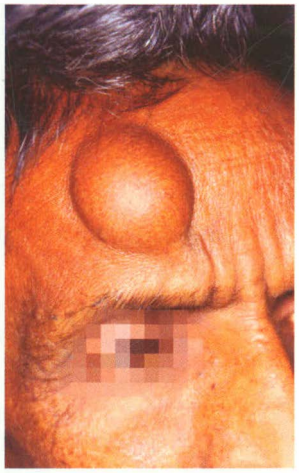


Fig. 15.9: Lipoma forehead. An undergraduate gave the diagnosis as dermoid cyst. You see the location. It is not the classical site of dermoid cyst and the 'slip sign' was positive. Even though face is not a common site, lipoma can occur in this location. Another differential diagnosis for this swelling is sebaceous cyst

- Commonly present as a subcutaneous swelling. It is ***freely mobile.*** The flank is the commonest site. Shoulder region, neck, back, upper limbs are the other common sites. (For various locations *see* Table 15.1, Figs 15.8 and 15.9.) Some lipomas from the chest wall can be of large size.
- ***Dimpling sign:*** **Fibrous bands connect a lipoma to the skin.** When the skin **moved,** a dimple appears **on the skin.**

2. Multiple lipomatosis (Fig. 15.6)

- Such lipomas are multiple and very often tender because of *nerve elements* mixed with them. Hence, they are called *multiple neurolipomatosis. Dercum's disease* is one example of this variety *(Adiposis dolorosa)* wherein tender, lipomatous swellings are present in the body, mainly the trunk.

3. Uncapsulated lipoma (diffuse)

- Diffuse variety is a rare type of lipoma. It is called pseudolipoma. It is an overgrowth of fat without a capsule.

Histological types of lipoma

1. **Fibrolipoma:** Since fibrous tissue is mixed with fat, lipoma feels hard.
2. **Neurolipoma:** *Painful* lipoma, because of presence of nerve elements.
3. **Naevolipoma:** Lipoma is usually relatively avascular but this variety is vascular.

Table 15.1 Various types of lipoma

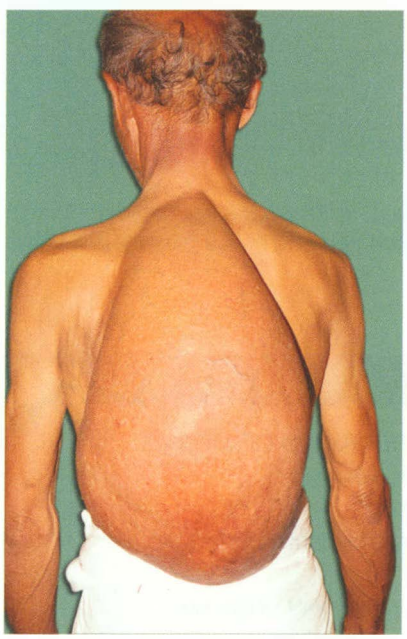
Location	Presentation	Differential diagnosis	Significance
1. Subcutaneous Shoulder Flank	Mobile Lobular **Edge slips under palpating fingers.**	Neurofibroma	**The most common variety**
2. Subfascial Limbs, palm, sole	Difficult to appreciate the edge and lobulation	Implantation dermoid, TB tenosynovitis	Subfascial lipoma of the scalp—erodes bone.
3. Subsynovial, intra-articular	Swelling in relation to knee joint, elbow joint (Figs 15.12 and 15.13)	Bursa Baker's cyst	Intra-articular lipomas are rare
4. Intermuscular Thigh Shoulder region	Swelling of the thigh. On contraction of the muscles, it becomes more firm due to transmitted pressure	Fibrosarcoma Haematoma	Chances of developing liposarcoma are more
5. Parosteal	Under the periosteum of bone, feels hard	Bony tumour	Very, very rare
6. Submucous Intestines, larynx	Asymptomatic or stridor or pain abdomen	Intestinal tumour Laryngeal tumour	Intussusception
7. Subserosal Retroperitoneum	Retroperitoneal swelling (Fig. 15.7)	Hydronephrosis Retroperitoneal cyst	Liposarcoma
8. Extradural	Very rare	–	–
9. Intraglandular	Breast, pancreas	Cystic lesions	Very rare

Treatment

1. An **incision** is given over the swelling. **Dissection** is carried out all around, separating it from underlying tissues and it is excised (Fig. 15.11).
2. **Small lipoma** can be removed by incising the skin followed by squeezing the lipoma out (no dissection method).

Complications

1. **Liposarcoma:** The current view is that lipomas are benign and do not turn into malignancy. However, a few retroperitoneal lipomas and lipoma in the thigh can turn into liposarcoma after many years of growth. Malignancy should be suspected when:
   - The swelling grows rapidly (Fig. 15.10).
   - It becomes painful due to infiltration of nerves.
   - The swelling becomes vascular and red coloured with dilated veins over the surface.
   - Surface is warm due to increased vascularity.
   - Skin fungation or fixation occurs later
   - Mobility gets restricted because of infiltration into deeper planes such as muscle.
   - Liposarcoma spreads via blood. It rarely spreads via lymphatics. Metastasis in the lung can rarely occur from liposarcoma producing multiple chest secondaries.
   - Liposarcoma is treated by wide excision followed by reconstruction either by split skin graft or by flaps. In the thigh, sometimes radical surgery may amount to compartmental excision. Chemotherapy and radiotherapy can also be used but the benefit is doubtful (*see* page 663).



Since 3 months, it is rapidly growing, observe dilated veins and shiny skin. There is local rise of temperature. These features are suggestive of sarcoma.

Fig. 15.10: Pedunculated lipoma of the back of 15 years duration with features suggestive of sarcomatous change

2. Calcification
3. Myxomatous degeneration: **Occurs only in retroperitoneal lipoma.**
4. **Intussusception**—due to submucosal lipoma of terminal ileum is an abdominal emergency.
5. Saponification (*see* Key Box 15.3 and Figs 15.12 and 15.13)

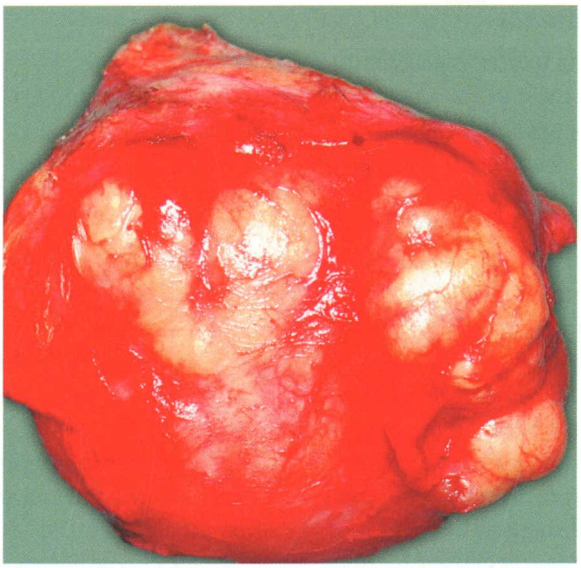


Fig. 15.11: Excised specimen of lipoma—see the lobularity

CLINICAL NOTES

A 32-year-old man presented with gross swelling of the right leg. He had seen two surgeons earlier who had told him that he had deep vein thrombosis but no treatment was offered. Examination revealed an obvious mass which was palpable in anteromedial and posterior compartment. MRI revealed an intermuscular mass. At exploration, an intermuscular lipoma weighing 700 g was excised (Figs 15.12 and 15.13).

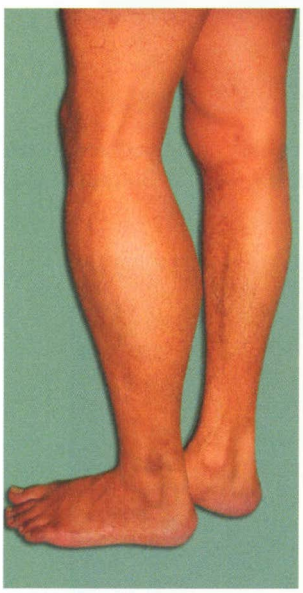


Fig. 15.12: Compare both the legs. Local gigantism can also be caused by extensive lipomatosis involving the leg. This was treated initially as filariasis

Fig. 15.13: Soleus muscle is cut, lipoma is seen coming out of the deeper plane. Luckily no major neurovascular bundle was involved. The patient had a smooth recovery

KEY BOX 15.3

SOME 'RARE' FACTS IN LIPOMA

- It is rare in children
- Rarely gives rise to transillumination (if size is big)
- Rarely gets infected (because it is relatively avascular)
- Diffuse variety is rare
- Neurolipomas are rare
- Rarely they turn into malignancy

NEURAL TUMOURS

NEUROMA

They are uncommon benign tumours which arise from sympathetic nervous system or spinal cord. They can be classified into true neuromas and false neuromas.

True neuroma

1. **Ganglioneuroma:** It consists of ganglion cells and nerve fibres of sympathetic chain. They are slow-growing tumours. When present in the neck as a *parapharyngeal mass,* it can cause dysphagia. These tumours can occur in the neck, retroperitoneum or mediastinum. Excision of the tumour is the treatment (*see* clinical notes).
2. **Neuroblastoma:** It consists of poorly differentiated cells. It occurs in young children. It is interesting to know that this tumour can undergo spontaneous regression.
3. **Myelinic neuroma:** It is very rare. It arises in relationship with spinal cord made up of myelinic fibres.

   - Does not contain any ganglion cells.

   All these three tumours are called *true neuromas.*

False neuroma

These tumours arise from the connective tissue of the sheath of nerve endings.

They occur following nerve injuries, lacerations or after amputation. They are of two types:

CLINICAL NOTES

An 18-year-old engineering student who had backache was examined by an orthopaedician and referred to general surgery. CT scan of the abdomen revealed mass in the paraspinal region in the retroperitoneum. Laparotomy and excision of the mass was done. It was a ganglioneuroma. Paraspinal region is one of the common sites of ganglioneuroma.

1. ***End-neuroma*** occurs after amputation due to proliferation of nerve fibres from the distal cut end of the nerve. This produces a bulbous swelling. If it is caught in the suture line or due to pressure of the prosthesis, it produces severe neuralgic pain. To avoid this, when an amputation is being done, the nerve is pulled downwards and cut as high as possible so that it retracts upwards (Fig. 15.14).
2. ***Lateral neuromas*** occur due to partial injury to the nerve on the lateral aspect (Fig. 15.15).

Treatment

Excision of the neuroma.

NEUROFIBROMA

It is a benign tumour arising from the connective tissue of the nerve sheath. Typically, it produces a *fusiform swelling* in the direction of the nerve fibres. The tumour contain both neural (ectodermal) and fibrous (mesodermal) elements.

Clinical types

1. Single subcutaneous neurofibroma (local)

Commonly affects the peripheral nerves such as ulnar nerve, median nerve or cutaneous nerves. Occurs in adults.

Clinical features

- **Presents as a painful, subcutaneous nodule.**
- Tingling and numbness, paraesthesia in the distribution of the nerve, specially when the nodule is compressed.
- Round to oval swelling in the direction of nerve fibre.
- Smooth surface, with round border. The swelling moves at right angles to the direction of nerve fibres. Vertical mobility is absent.
- Consistency is firm. Sometimes, it is hard.
- Being a subcutaneous swelling, the skin can be lifted up.

Differential diagnosis of subcutaneous nodule

1. Neurofibroma, 2. Metastatic nodule, 3. Cutaneous T cell lymphoma, 4. Chronic abscess

Fig. 15.14: End neuroma

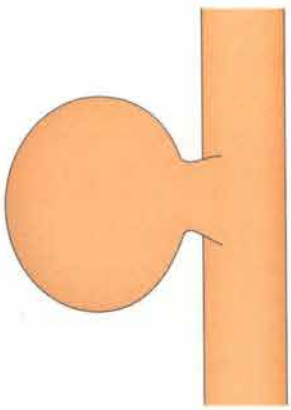


Fig. 15.15: Lateral neuroma

Treatment

- It is treated by excision.
- In most of the cases, excision is easy as the tumour is we encapsulated.

2. Generalised neurofibromatosis: von Reckling hausen's (vR) disease (Type I) (Figs 15.16 to 15.18)

- This is an autosomal dominant disorder transmitted by bot sexes. The whole body is studded with cutaneous nodule of varying sizes. They are soft and nontender.
- Coffee brown pigmentation is characteristic of this conditio (*café au lait* spots, Fig. 21.). *Café au lait* spots can b associated with involvement of cranial nerves—VIIIth nerv (auditory nerve) *acoustic neuroma—**a cerebellopontin angle tumour. Popularly called vestibular schwannoma.***
- Fibroepithelial skin tags are often present.
- Type I is caused by ***gene mutation on chromosome 17.***
- The presence of skin pigmentation is an indication of th common neuroectodermal origin of nerve sheath cells an melanocytes.
- Skeletal deformities such as kyphoscoliosis or osteoporosi are common.

Fig. 15.16: von Recklinghausen's with plexiform neurofibromatosis (*Courtesy:* Dr Prashanth Shetty, Professor, Dept. of Surgery, KMC, Manipal)

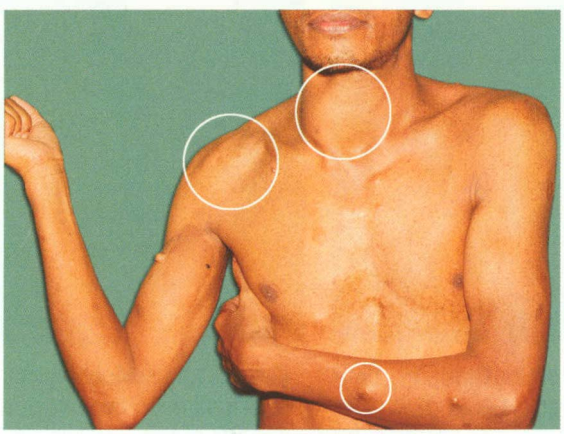


Fig. 15.17: Case of vR disease with multiple nodules, pigmentation and schwannoma of vagus nerve (*Courtesy:* Dr Siddarth Bhandary, Professor, Dept. of Surgery, KMC, Manipal)

- It may be associated with phaeochromocytoma (high blood pressure).
- Sarcomatous changes do occur

PEARLS OF WISDOM

Record the blood pressure in all cases of von Recklinghausen's disease because it may be associated with phaeochromocytoma.

Bilateral vestibular schwannoma or acoustic neuromas are pathognomonic of neurofibromatosis type 2, a syndrome resulting from chromosome 22 mutation. It is also associated with increased incidence of meningiomas and gliomas.

3. Plexiform neurofibromatosis (Trigeminal) (Fig. 15.19)

- In this condition, the branches of 5th cranial nerve are commonly affected. It can also involve the peripheries.
- The affected part is grossly thickened due to **fibromyxomatous degeneration**.
- When it involves the branches of trigeminal nerve, following problems can occur:
  - **Tingling paraesthesia** in the distribution of Vth nerve, especially ophthalmic division.
  - When it attains a huge size, it can **obstruct the vision**. As it grows bigger in size, it hangs in front of the neck, as a grossly thickened pendulous fold of skin.
- **Treatment:** Very difficult. Excision can be attempted with plastic surgery repair.

4. Elephantiasis neuromatosa

- This condition affects the limbs. It represents an advanced stage of plexiform variety. Gross thickening of subcutaneous tissue gives the appearance of elephant's leg. The skin is dry and coarse (Fig. 15.20).

Complications of neural tumours (Key Box 15.4)

KEY BOX 15.4

COMPLICATIONS OF NEURAL TUMOURS

- Atrophy of muscles
- Dumb-bell tumours from dorsal spinal nerve root can cause backache or paralysis
- Acoustic neuroma—Deafness
- Cystic degeneration
- Sarcomatous change

5. Pachydermatocoele

- This refers to the plexiform lesions mainly found in the neck as a thickened, coiled single mass.

NEURILEMMOMA (SCHWANNOMA)

- This is a benign tumour arising from ***Schwann cells.***
- Commonest site is the ***acoustic nerve.*** However, vagus nerve is the most common peripheral site (Fig. 15.22).
- They can be single or multiple and present with a fusiform swelling in relationship with the nerves.
- They can also arise from a peripheral nerve. ***Sensory branches*** are affected more frequently (Table 15.2).
- They can also be seen in mediastinum and retroperitoneum.
- They are ***soft, lobulated, well encapsulated tumours.***
- They are ***benign*** and ***do not turn into malignancy.***

Fig. 15.18: von Recklinghausen's disease—Tumours are multiple, congenital, familial

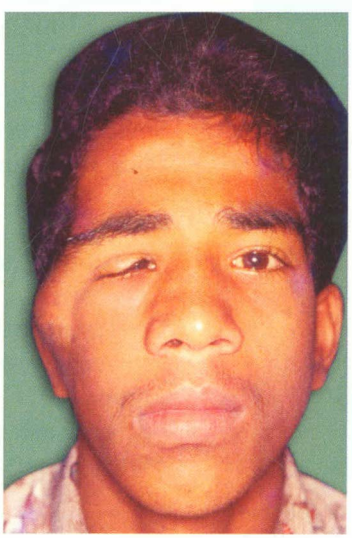


Fig. 15.19: Plexiform neurofibromatosis (*Courtesy:* Dr Rohit Jain, Asst. Professor, Department of Surgery, KMC, Manipal)

Fig. 15.20: Elephantiasis neuromatosa (*Courtesy:* Dr Balakrishna Shetty, Prof of Surgery, KS Hegde Medical Academy, Mangalore)

Table 15.2 Comparison between neurofibroma and schwannoma

Neurofibroma	Schwannoma
• More common	• Less common
• Ectodermal and mesodermal origin	• Ectodermal origin
• Subcutaneous (forearm) nerves are the commonest site	• Acoustic nerve is the commonest site
• Multiple lesions are common	• Very rare
• Feels firm or hard	• Soft
• Tender	• Nontender
• Can turn into sarcoma	• **Does not turn into sarcoma**
• Often nerve fibres are entangled with tumour. Hence, excision involves sacrificing nerve also	• Well encapsulated. Hence, **enucleation is possible** without sacrificing nerve

Fig. 15.21: The *café au lait* spots. More than five such spots will appear by early life (*Courtesy:* Dr Prashanth Shetty, Professor, KMC, Manipal)

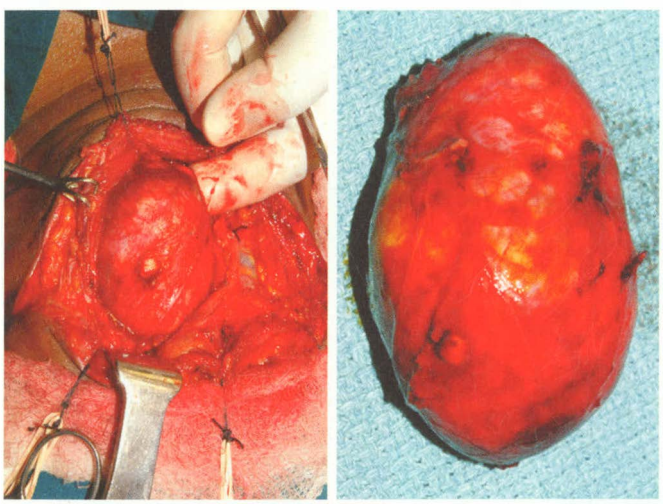


Fig. 15.22: Vagal schwannoma at surgery—you can see the displaced carotid artery and internal jugular vein. Second picture showing the specimen (*Courtesy:* Prof Balakrishnan, Dept. of ENT, Kasturba Hospital, Manipal. Initially it was diagnosed as solitary nodule of the thyroid gland but it was not moving with deglutition)

Treatment

- Excision of the tumour can be done without sacrificing the nerves because the tumour is well encapsulated and displaces the nerve.

HAMARTOMA

- It is a tumour-like developmental malformation of the tissues of a particular part of the body wherein it is arranged haphazardly.
- Hamartoma is a Greek word which means **fault** or **misfire**. It is not a clinical diagnosis.

A few examples of hamartoma

Haemangioma, neurofibroma, glomus tumour, benign naevus, lymphangioma.

Characteristic features

- Being a developmental anomaly, they are seen at birth or in early childhood.
- In adults, there is a long history of swelling.
- **Being a malformation (not a tumour), it does not have a capsule.**
- They can be single or multiple
- Some may **regress** as in **strawberry angioma**.
- They are benign lesions.

Treatment

- **Excision** is not only curative but also gives a correct diagnosis.
- Care should be taken when it contains vascular tissue such as haemangioma or neural tissue as in cases of neurofibroma.
- Facial nerve and its branches may be damaged while excising hamartomatous lesions over the face.

CHORDOMA

- Rare tumour
- Remnant of notochord (origin)
- Sacrococcygeal region (common site)

- Resection is difficult, chances of neurological deficit and bleeding are high.
- Radioresistant

MALIGNANT TUMOURS

Types of malignant tumours

- They are of two types: Carcinoma and sarcoma. Carcinoma arises from epithelium–ectodermal, endodermal or mesodermal in origin.
- Sarcomas arise from soft tissues or bone and are derived from mesoblast or mesenchymal tissues.
- It may be observed that mesoderm can give rise to carcinoma and mesenchymal sarcoma also.

Pathology (Table 15.3)

Spread

1. **Local spread:** Generally, local spread occurs into adjacent structures. Few examples are given:
   - Carcinoma cheek—fixity to mandible
     Significance: May necessitate removal of mandible along with wide excision (Fig. 15.23A).
   - Squamous cell carcinoma—fixed to tibia may necessitate an amputation.
2. **Lymphatic spread:** It is one of the most important features of **carcinoma** (Key Box 15.5). As you complete reading this book, you will come across many cases and many examples of lymphatic spread of malignant tumours. A few sarcomas also spread by lymphatics (*see* page 225). Different types of lymphatic spread are given below:
   - **Embolisation:** More aggressive tumour means more aggressive spread—by embolisation wherein nodes can be enlarged in a far away station, e.g. malignant melanoma (Fig. 15.23B).
   - **Permeation:** Refers to tumour cells travelling along the lymphatic vessel, e.g. carcinoma tongue with submandibular node enlargement (Fig. 15.23C).

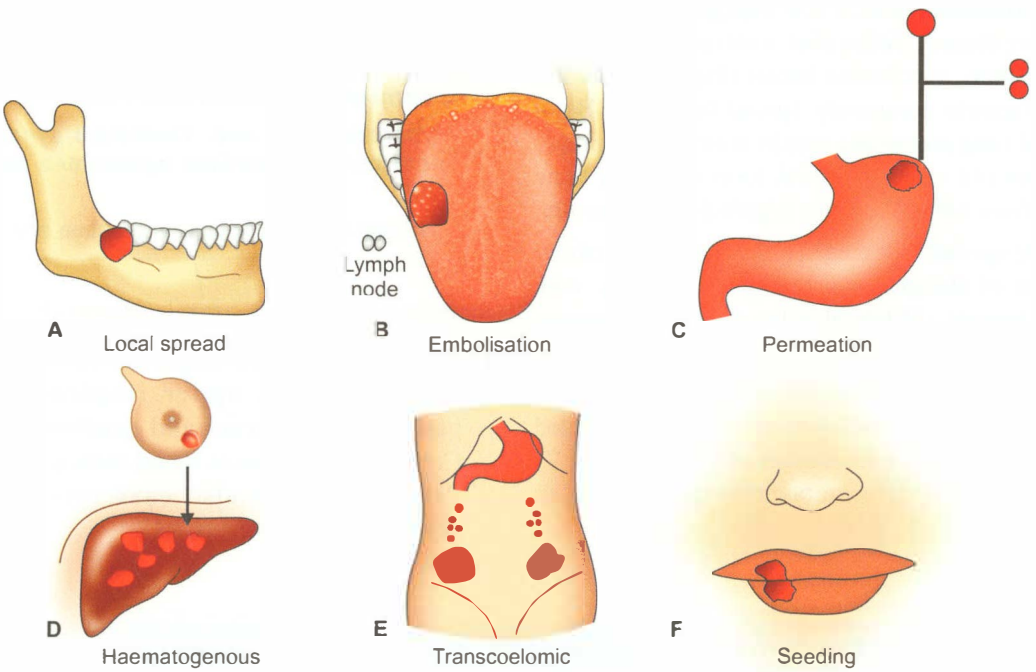


Fig. 15.23: Spread of malignant tumour (*see* text)

Table 15.3 Pathology of tumours

Terminology	Explanation	Examples
1. **Well differentiated**	Cells that resemble very closely their normal counterparts	Well-differentiated squamous cell carcinoma
2. **Undifferentiated** (anaplasia)	Loss of structural and functional differentiation	Poorly-differentiated carcinoma (anaplasia)
3. **Dysplasia**	Loss in the uniformity, loss in the architectural orientation	Barrett's columnar cell lined oesophagus
4. **Carcinoma *in situ***	Dysplastic changes involving **entire thickness of the epithelium**	Cheek, tongue, breast, etc.
5. **Apoptosis**	Programmed cell death	Seen in malignant tumours

KEY BOX 15.5

CARCINOMA

ORIGIN

- Ectodermal—skin cancer
- Endodermal—gut cancer
- Mesodermal—renal carcinoma

TYPES

- Squamous cell carcinoma
- Basal cell carcinoma
- Glandular

- **Retrograde lymphatic spread:** When main lymphatic pathway is blocked retrograde spread can occur and a node in an unusual location may get enlarged, e.g. **Irish node (left axillary node enlargement in carcinoma stomach).**

3. **Haematogenous spread:** This is most important method of spread of **sarcomas**. Also, a few malignancies such as renal cell carcinoma, follicular carcinoma thyroid, carcinoma prostate, carcinoma breast (Fig.15.23D) and malignant melanoma commonly spread by blood. Bony metastasis and lung metastasis results from blood spread. Bone metastasis can vary from mild form with only bone pain to severe form with quadriplegia/pathological fracture.
4. **Transcoelomic spread:** Spread through peritoneal cavity by dislodgment of malignant cells, e.g. Ca stomach with Krukenberg's tumour—Bilateral bulky ovarian metastasis, commonly seen in premenopausal patients (Fig. 15.23E).
5. **Seeding** (Fig. 15.23F): A few examples are given below:
   - Cancer of lower lip spreading to upper lip, also called kiss cancer. Other example: Cancer of vulva.
   - Incision and 'port' site metastasis (port refers to laparoscopic port).

Comparison of benign and malignant tumours is shown i Table 15.4.

PARANEOPLASTIC SYNDROMES (PNS)

These are interesting syndromes and are listed i Table 15.5.

Certain cancers produce some specific clinical syndrome (symptom complexes other than cachexia) which cannot b explained by their local or distant spread or by the hormone produced by the tissue of origin of these tumours. These ar called Paraneoplastic syndromes (Key Box 15.6).

Just to give an example, hypercalcaemia due to skeleta metastasis from carcinoma breast is not considered as PNS but if it occurs without skeletal metastasis, it is considered a PNS.

KEY BOX 15.6

IMPORTANT FEATURES OF PNS

- Incidence : 10–15% of patients with cancer
- It may be the earliest manifestation (primary can be occult)
- **Bronchogenic cancer and breast cancer are most commonly associated with PNS.**
- **Hypercalcaemia and Cushing's syndromes are the most common clinical syndromes associated with PNS.**
- PNS can be a major clinical problem and can be treated.

Aetiology of carcinoma in general

1. **Tobacco** is the most important factor in the development of **lung cancer, upper respiratory tract cancer, gastrointestinal tract and genitourinary tract cancer.** Carcinoma pancreas is found more commonly in smokers. Passive smokers also have increased incidence of development of cancers.

Table 15.4 Comparison of benign tumours and malignant tumours

Feature	Benign	Malignant
Growth	Very slow	Rapid
Duration	Long	Short
Pain	Usually not a feature	Pain can be present due to local infiltration
Mobility	Present	Restricted
Fixity	No	Can be present
Consistency	Firm/soft	Hard, irregular
Spread	No	Spreads
Capsule	Capsulated	Uncapsulated
Recurrence after surgery	Does not occur	Can occur if wide excision is **not** done

Table 15.5 Paraneoplastic syndromes

Clinical syndrome	Underlying major cancer	Mechanism
1. Endocrinopathies		
• Cushing's syndrome	Small cell Ca lung, Ca pancreas	ACTH or ACTH-like substance
• Hyponatraemia	Small cell Ca lung	ADH or atrial natriuretic factor
• Hypoglycaemia	Hepatoma	Insulin or insulin-like substance
• Hypercalcaemia	Squamous cell Ca lung Ca breast, Ca kidney	PTH like substance
• Carcinoid syndrome	Bronchial carcinoid, Ca pancreas, Ca stomach	Serotonin, bradykinin
• Polycythaemia	Renal cell Ca Cerebellar haemangioma	Erythropoietin
2. Muscle syndrome		
• Myasthenia gravis	Thymoma	Immunologic
3. Bone and soft tissues		
• Hypertrophic osteoarthropathy and clubbing of fingers	Adenocarcinoma lung	Unknown
4. Vascular		
• Venous thrombosis	Carcinoma pancreas	Hypercoagulability
• Disseminated intravascular coagulation	Ca pancreas, lung, prostate	Tumour products

Ca = Carcinoma

2. **Alcohol:** Smoking with alcohol increases the permeability of the upper digestive tract mucosa and respiratory mucosa to the carcinogens. Thus, they increase the incidence of cancer. Hepatocellular cancer is commonly found in alcoholic cirrhotic liver.
3. **Ionising radiation:** Atomic bomb blasts in Japan have definitely resulted in increased number of cases of breast cancer in premenopausal women and leukaemia in children.
4. **Ultraviolet radiation: Causes all types of skin cancers.**
5. **Genetic causes** (Key Box 15.7)
6. **Hereditary causes**
   - **MEN** syndrome: Medullary carcinoma thyroid (**M**ultiple **E**ndocrine **N**eoplasia)
   - **FPC**: Colonic cancer (Multiple) (**F**amilial **P**olyposis **C**oli)

KEY BOX 15.7

GENETIC / DEFECTIVE DNA REPAIR

1. Xeroderma pigmentosa	:	Skin cancer
2. Bloom's syndrome	:	Acute leukaemia, various cancers
3. Fanconi's anaemia	:	Acute leukaemia, squamous cell carcinoma, hepatoma
4. Ataxia-telangiectasia	:	Acute leukaemia, lymphoma, breast cancer

   - Li-Fraumeni syndrome: Familial breast cancer.
   - Retinoblastoma.
7. **Dietary factors**
   - **Red meat:** Carcinoma colon, carcinoma breast.
   - **Fat:** Carcinoma breast, carcinoma colon.
   - **Smoked, charred fish:** Carcinoma oesophagus, carcinoma stomach.
8. **Chemicals**
   - Benzanthracenes: Skin cancer when painted on the skin
   - Benzopyrenes: Lung cancer
   - β-naphthylamine: Bladder cancer
   - Nitrosamines and amides: Cancer stomach
   - Aflatoxin B: Hepatocellular carcinoma
   - Asbestos: Lung cancer
9. **Viral factors**
   - Human T-cell leukaemia virus type 1 (HTLV-1): T cell leukaemia/lymphoma. (RNA virus)
   - Human papillomavirus (HPV) Cancer cervix, cancer urogenital region
   - Epstein-Barr virus: Burkitt's lymphoma.

SOFT TISSUE SARCOMAS (STS)

These are malignant tumours arising from soft tissues. Thus, they can occur in any part of the body. Examples are given in Table 15.6.

Table 15.6 Soft tissue sarcoma

Tissue of origin	Name
• Mesenchymal tissue	Malignant fibrous histiocytoma (MFH)
• Adipose	Liposarcoma
• Smooth muscle	Leiomyosarcoma (GIST)
• Striated muscle	Rhabdomyosarcoma
• Synovial tissue	Synovial sarcoma
• Neural tissue	Malignant schwannoma
• Uncertain	Epithelioid sarcoma
• Blood vessels	Angiosarcoma
• Lymph vessels	Lymphangiosarcoma

Introduction

- These are malignant tumours which are **fatal if untreated** or mistreated. Most of them occur in young patients as painless lumps. CT scan, MRI and incision biopsy (details later) are key investigations. Early diagnosis and curative resection have a major role in the management of soft tissue sarcomas. In addition to TNM staging, pathological grading of the tumour has been included **GTNM staging**.

Aetiology/epidemiology of STS (Key Box 15.8)

1. **Genetic factors: Genetic mutations/gene rearrangements** have been implicated in the pathogenesis of STS. Two genes that are most relevant to soft tissue tumours are retinoblastoma **(Rb) tumour suppressor gene and p53 tumour suppressor gene.** Various familial syndromes are associated with STS such as Li-Fraumeni syndrome, Gardner's syndrome and von Recklinghausen's disease. Several oncogenes have been identified.
2. **Exposure to radiation:** 8 to 50-fold increase in the incidence of STS is reported in patients who are treated for cancer of the breast, cervix and ovary by radiation.
3. **Neurofibromatosis type I:** Specially mentioned here because it is the only benign tumour progressing to STS.
4. **Exposure to chemicals:** Thorium oxide, vinyl chloride, arsenic have been implicated in hepatic angiosarcomas.
5. **Trauma:** Chronic tissue trauma is blamed as a triggering factor for development of STS. Others believe that trauma draws attention of the patient to the STS.
6. **Immunosuppression:** Kaposi's sarcoma occurs in HIV-AIDS patients.
7. **Chronic lymphoedema** can give rise to lymphangiosarcoma. Postmastectomy lymphangiosarcoma is called Stewart-Treves syndrome.

Clinical features

Sarcomas are rapidly growing vascular growth (Table 15.7).

KEY BOX 15.8

AETIOLOGY OF STS—SUMMARY

Genetic factors
Exposure to radiation
Neurofibromatosis (NF-1)
Exposure to chemicals
Trauma
Immunodeficiency
Chronic lymphoedema
Remember as **GENETIC**

TNM STAGING Staging of soft tissue sarcoma

1. Grade (G)
- G1 – Well differentiated
- G2 – Moderately differentiated
- G3 – Poorly differentiated
- G4 – Undifferentiated

2. Primary Tumour (T)
- a: Superficial tumour
- b: Deep tumour
- T1: Tumour < 4–5 cm in greatest dimension
- T2: Tumour > 5 cm in greatest dimension

3. Regional lymph nodes (N)
- N0 : No nodal metastasis
- N1 : Regional lymph node metastasis present.

4. Distant Metastasis (M)
- M0 : No distant metastasis
- M1 : Distant metastasis present
- G – TNM staging system

Stage grouping

I	G1	T1–T2	N0	M0
II	G2	T1–T2	N0	M0
III	G3	T1–T2	N0	M0
IV	G1–G3	T1–T2	N1	M0
	G1–G3	T1–T2	N1	M1

Investigations/diagnostic imaging

- **Routine blood tests**
- **Chest radiography:** Presence of cannon ball metastasis alters the staging, treatment policy and prognosis.

PEARLS OF WISDOM

If STS is more than 5 cm (T2), computed tomography (CT) of the chest should be considered.

- **CT scan** is useful in evaluating retroperitoneal sarcomas. It can define structures, infiltration into neighbouring structures, hydronephrosis, etc. CT can guide a ***core biopsy also***.

Table 15.7 Comparison of carcinoma and sarcoma

	Carcinoma	Sarcoma
1. Cell of origin	Ectodermal or endodermal (epithelial)	Mesodermal (mesenchymal)
2. Age group	Elderly, 40–60 years	Young, 10–30 years
3. Rate of growth	Slow	Fast
4. Presentation	**Nonhealing ulcer,** cauliflower-like growth with everted edges and induration. Fixity is a late feature	**Fleshy mass, red and vascular, dilated veins** over the surface, local rise of temperature. Ulceration is a late feature
5. Spread	**Lymphatic spread** is very common, both by emboli and permeation. Blood spread does occur as in renal cell carcinoma, thyroid carcinoma, breast carcinoma, etc.	**Blood spread** occurs very early and results in cannon ball secondaries in lung. Rhabdomyosarcoma, synovial sarcoma, epithelioid sarcoma. Malignant fibrous histiocytoma, angiosarcoma spread by lymphatics
6. Microscopy	**Cell nests or epithelial pearls** are seen in well-differentiated cancers	Malignant cells resemble their cell of origin. Thus, **spindle-shaped cells** are found in **fibrosarcoma**
7. Treatment	**Surgery** is the main treatment of ectodermal cancers, surgery or radiotherapy for ectodermal lesions. Chemotherapy is not very useful	**Surgery** is the main modality of treatment. Radiotherapy and chemotherapy are also beneficial when the tumour is more than 5 cm

- **MRI** is the investigation of choice when STS occurs in **extremities to delineate muscle groups,** bones, vascular structures, etc.
- **Biopsy: FNA**
  - Acceptable first investigation to prove the diagnosis.
  - Useful to detect metastatic disease
  - To detect local recurrence
  - Ideal for superficial lesions
  - **Disadvantage:** Cannot assess tumour grading. Tissue is not sufficient for diagnostic tests.
- **Core needle biopsy**
  - Safe and accurate
  - Tissue is sufficient for grading, electron microscopy and flow cytometry
  - With CT guidance, core biopsy can be taken from deeper structures also.
- **Incisional biopsy**
  - When core biopsy tissue is not adequate, incisional biopsy is indicated (Key Box 15.9).

KEY BOX 15.9

GUIDELINES WHILE DOING AN INCISIONAL BIOPSY

- Incision should be oriented longitudinally in STS of extremities to facilitate removal of biopsy site, scar and tumour *en bloc*
- Flaps should not be raised
- Perfect haemostasis should be achieved
- Prevent dissemination
- Avoid drain

Treatment (*see* next page)

- The aim is to achieve local control and to treat metastasis including subclinical metastasis, thus trying for a cure.
- **Surgery is the first line of treatment** varying from a wide local excision to amputation or disarticulation (5%), when it occurs in the extremities. **Low-grade tumours** can be treated on **2 cm wide excision** of surrounding normal tissue and **high-grade tumours by 4 cm margin** (Key Box 15.10).
- Small tumours less than 5 cm have not been associated with recurrence. Hence, radiotherapy may not be required **but if grade is high, RT is required** (Key Box 15.11).
- Tumours do respond to radiotherapy and chemotherapy.

Role of chemotherapy

- High-grade tumours have high potential of metastasis. Hence, combination chemotherapy is to be considered before or after surgery.
- The most favoured combination chemotherapy drugs include Mesna, Adriamycin, Ifosfamide and Dacarbazine (MAID).
- The success rate is around 10–20%.

Sarcomas that metastasise to lymph nodes*

- **R**habdomyosarcoma
- **A**ngiosarcoma
- **C**lear cell carcinoma
- **E**pithelioid sarcoma
- **S**ynovial sarcoma

Remember as* **RACES

A FEW SURGICAL PROCEDURES DONE FOR SOFT TISSUE SARCOMA (Figs 15.24 to 15.29)

Fig. 15.24: A 24-year-old lady underwent excision of a swelling diagnosed as lipoma in a peripheral hospital which was reported as sarcoma

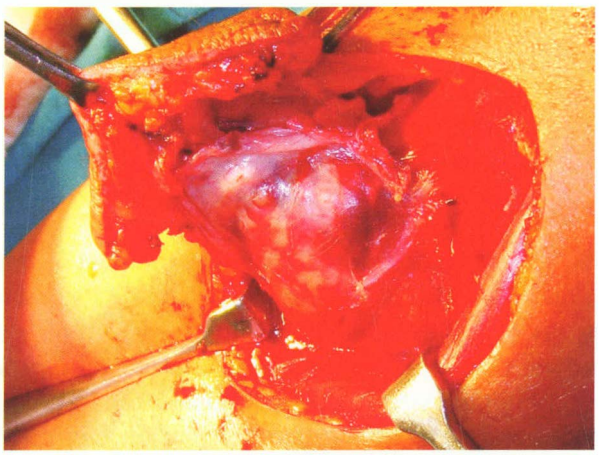


Fig. 15.25: The swelling was explored—as you can see it was infiltrating the muscles—the first surgeon had not excised the swelling completely

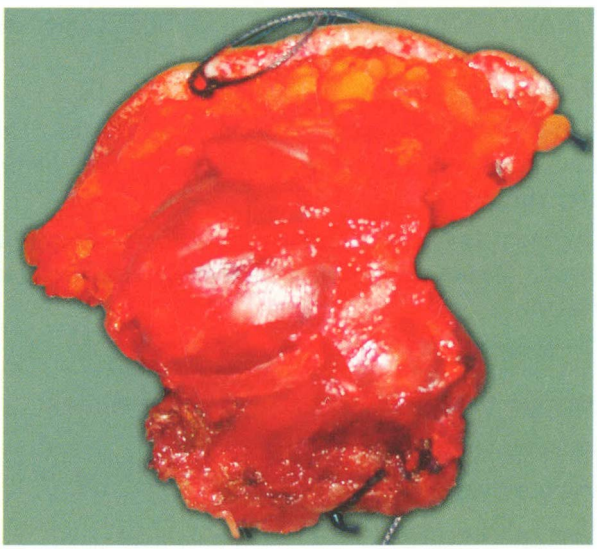


Fig. 15.26: Excised specimen showing skin, tumour and deeper muscle fibres. STS was 3 cm in diameter. Histopathology reported as synovial cell sarcoma. Margins were negative (*Courtesy:* Dr Ankur Sharma, Asst Professor, Dept. of Surgery, KMC, Manipal)

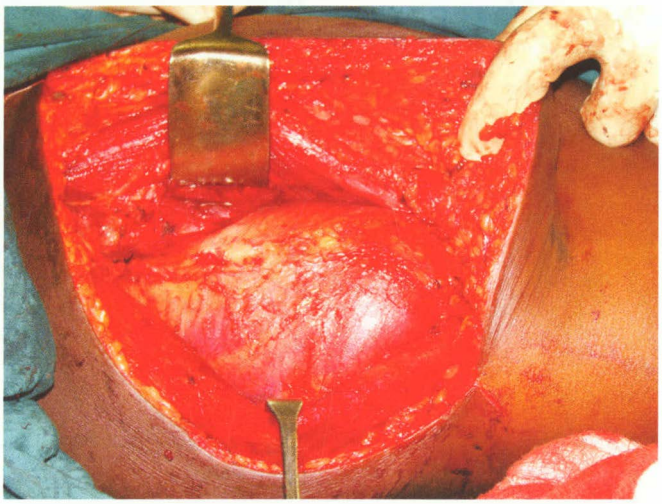


Fig. 15.27: A large (8 cm) liposarcoma of the thigh being explorec with a longitudinal incision of the thigh. He had undergone an incision biopsy earlier. The biopsy site is removed along with the skin

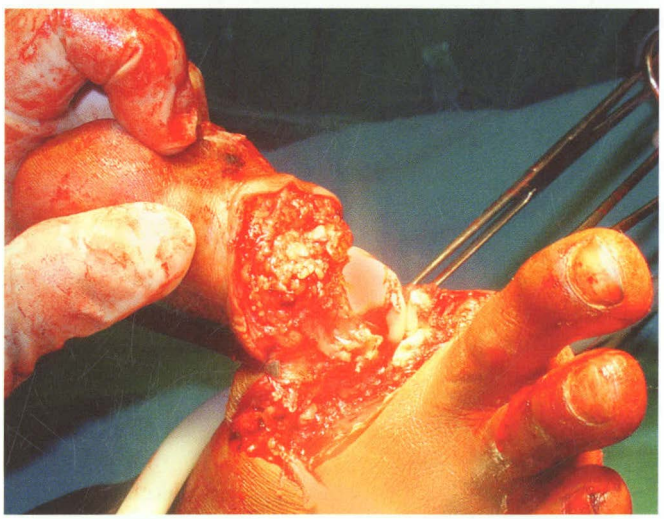


Fig. 15.28: Synovial sarcoma of the toe—disarticulation being done—highly malignant sarcoma

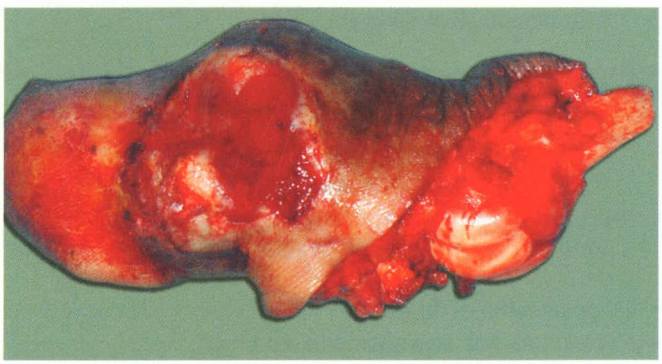


Fig. 15.29: Specimen of the growth along with the toe: Wide excision. No nodes were palpable in the leg

KEY BOX 15.10

SURGERY FOR SOFT TISSUE SARCOMA

- **Surgery is the primary and most effective therapy**
- Local wide excision with 2 cm to 4 cm of surrounding normal tissue should be removed. **2 cm for low grade and 4 cm for high grade tumour.**
- **Not to dissect along pseudocapsule** which is composed of tumour cells.
- If necessary, **excision should include nerves** and vessels followed by nerve grafts and arterial reconstruction.
- **STS rarely involve bones** and skin. Hence, wide resections of these structures are infrequently necessary.
- It should also include **previous scars.**
- If amputation can be avoided by giving preoperative radiotherapy, it is preferred first. It is then followed by wide excision/compartmental excision and postoperative radiotherapy. Dose of pre- and postoperative radiotherapy is 50–60 Gy given in 25 fractions. Brachytherapy is also given.
- **Margin negative resection should be the aim.**

KEY BOX 15.11

RADIOTHERAPY

- *It is not the first modality of treatment in STS.*
- Radiotherapy is given after margin negative resection.
- Surgery + RT has resulted in local control rates up to or more than 90%.
- External beam radiation therapy (electrons, protons or neutrons) is given with a margin of 5 to 7 cm or even more depending on size of tumour. Dose is 50 Gy given in 25 fractions—as preoperative dose.
- Dose is 60 to 70 Gy for postoperative treatment.
- In brachytherapy, multiple catheters are placed in tumour resection bed. Catheters are loaded with seeds containing Iridium—192.
- Dose of brachytherapy is 42–45 Gy to tumour bed over 4–6 days.
- Short period of treatment time and less systemic toxicity are advantages of brachytherapy.

Prognosis

- Almost 80% of STS metastasise to lungs within 2–3 years of the diagnosis.
- Prognosis depends on metastatic STS, grade of the tumour, size of the tumour, margins after resection and anatomical location.
- If pulmonary metastasis is resectable, 30% survival may be expected at 3 years.

DIFFERENTIAL DIAGNOSIS OF SOFT TISSUE SARCOMA

LIPOSARCOMA (Figs 15.36 and 15.37)

- It is a malignant fatty tumour (*see* page 550).
- **Common sites:** Proximal extremity, trunk or retroperitoneum (Fig. 15.30).
- They are generally large at the time of diagnosis, e.g. retroperitoneum. It results in gross swelling, which is firm to hard (more than 50% will be of > 20 cm size) (Key Box 15.12)

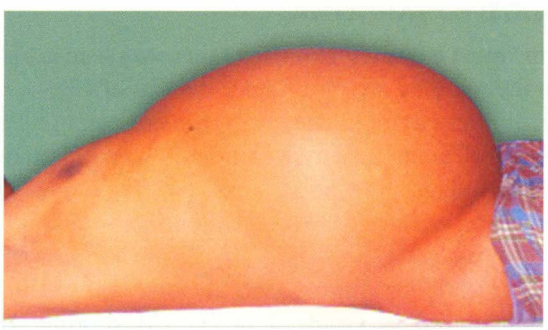


Fig. 15.30: Retroperitoneal liposarcoma

- The compression on blood vessels may result in oedema of the limbs when it occurs in retroperitoneum.
- Well-differentiated myxoid liposarcomas **are notoriously known to recur** many times before spreading to lungs. **Hence, prognosis is good** (Fig. 15.42).
- Pleomorphic and lipoblastic liposarcomas tend to be of higher grade and often present with metastasis.
- Wide excision/surgery is the primary treatment.
- They do respond to radiotherapy.

KEY BOX 15.12

RETROPERITONEAL SARCOMA (RPS)

- Most common RPS is liposarcoma or leiomyosarcoma.
- They constitute 15% of adult soft tissue sarcomas.
- May present as large masses of more than 20 cm at the time of presentation.
- Local recurrence and intra-abdominal spread is more common.
- Progressive abdominal distension, pedal oedema, young age and firm to hard retroperitoneal mass clinches the diagnosis.
- CT scan of chest and abdomen followed by FNAC/True cut biopsy for histology/grade of the tumour.
- Margin negative—**complete surgical resection** is the treatment of choice.
- Chemotherapy **has not been effective** against RPS.

MALIGNANT FIBROUS HISTIOCYTOMA (MFH)

- It is a malignant tumour of mesenchymal tissue (fibrous tissue). This is the recent nomenclature of sarcoma.

Fibrosarcoma or pleomorphic rhabdomyosarcomas are included under this. **Most of the so-called fibrosarcomas are presently included under MFH** (Fig. 15.31).

- These are **high-grade tumours** that lack differentiation.
- It can also arise from bone.
- **The MFH: Superficial type rarely metastasises and carries good prognosis.**
- Locations: Retroperitoneum, trunks and limbs (inter-muscular septae of adductors, scapulohumeral and pectoral muscles).

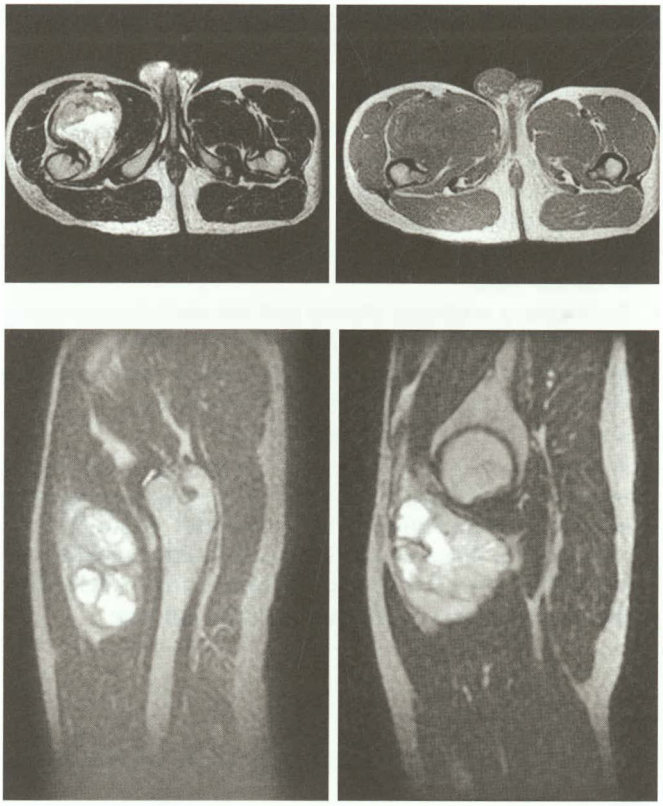
Clinical features

- Common in elderly patients (50 years) but can occur at any age.
- Slow-growing, firm to hard mass with restricted mobility.
- As the tumour is locally invasive, it infiltrates the muscles and adjacent structures. Thus, it can cause muscle weakness or pain, etc.
- **Spread:** Local spread is common. Distant metastasis by blood is late (lungs). Lymph node metastasis is rare.
- Like other sarcomas, dilated veins, local rise of temperature, restricted mobility and hardness will clinch the diagnosis.
- **MRI is the investigation of choice to know the extent of the disease** (Figs 15.32 to 15.35).
- Margin negative surgery should be the aim.

SYNOVIAL SARCOMA (Fig. 15.40)

- Any rapidly growing tumour in the region of joint or near the tendons in young patients (20–40 years), synovial sarcoma is to be considered.
- Common site: Shoulder, wrist, knee, etc.
- Age: Young between 20 and 40 years.
- Clinical features are similar to the other sarcomas—hard, painful mass.
- In addition to the local and blood spread, **it also spreads by lymphatic route**.

Fig. 15.31: Recurrent fibrosarcoma (MFH)



Figs 15.32 to 15.35: T1- and T2-weighted (W) axial MRI. Isointense to hyperintense mass lesion noted in the anterior compartment of right thigh. Sagittal T2-W MRI shows supero-inferior extent of the lesion

- Plain X-ray: It may show characteristic calcification.
- It is aggressive, with high rates of recurrence.
- **In the G-TNM staging system, they are grade 3.**

ANGIOSARCOMA (Fig. 15.41)

- 1 to 2% of soft tissue sarcomas.
- Affects elderly patients.
- They are high grade and aggressive tumours.
- They arise from skin and subcutaneous tissue rather than deeper tissues.
- Most of them occur in the **head and neck, breast and liver**.
- Surgery (excision) followed by radiotherapy/combination chemotherapy may have to be given.

RHABDOMYOSARCOMA

- It is the **most common soft tissue sarcoma** seen in children, even though they are rare (under the age of 15).
- It arises from **striated muscle**—painless enlarging mass.
- Resection/Chemotherapy/Radiotherapy (combination) is tried depending on location.
- **Sites:** Head and neck (30%), genitourinary system (25%), extremities (20%).
- All three varieties: Embryonal, alveolar and pleomorphic are considered as Grade 3 in GTNM staging. Hence, prognosis is not good (Fig. 15.43).

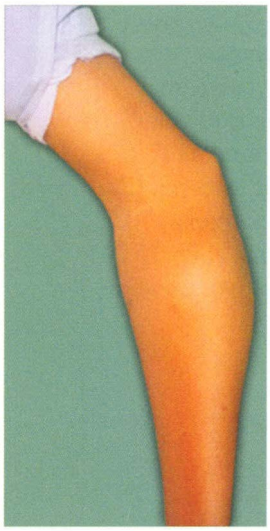


Fig. 15.36: Early stage of liposarcoma—this swelling had local rise of temperature and restricted mobility

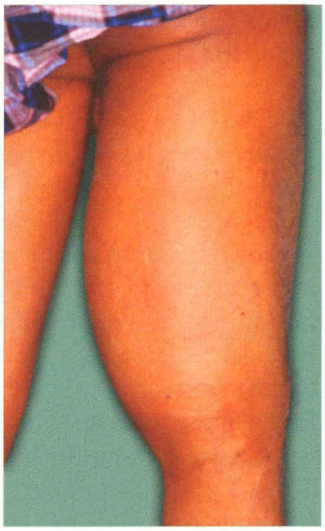


Fig. 15.37: Advanced case of liposarcoma—the swelling was hard and fixed. Dilated veins over the surface and location were characteristic

Fig. 15.38: Swelling in the elbow region for two years duration presented to the hospital with bleeding and fungation—advanced case of epithelioid sarcoma

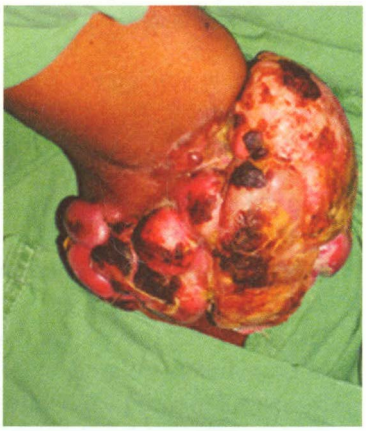


Fig. 15.39: Ulcerated lesion in the elbow region of two years duration presented to the hospital with bleeding—a case of dermatofibrosarcoma protuberans

Fig. 15.40: Synovial sarcoma left shoulder region. *See* the secondary varicosity due to pressure effects. Nonextremity sarcomas have poor prognosis

Fig. 15.41: Bleeding vascular lesion in the ankle region—angiosarcoma (*Courtesy:* Dr Mallikarjuna Desai, HOD, Surgery, SDMC, Dharwar, Karnataka)

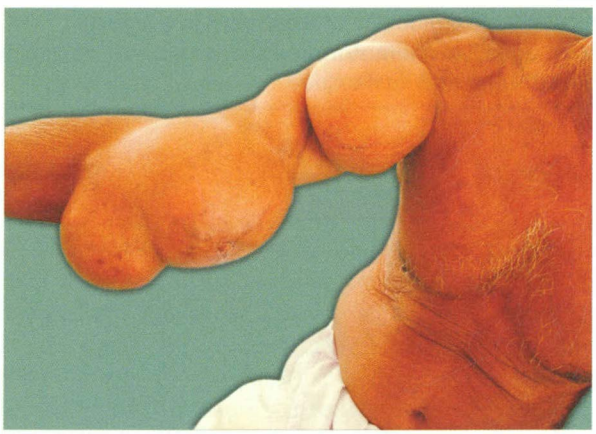


Fig. 15.42: Six times recurrent myxoid liposarcoma since 8 years. It has good prognosis (*Courtesy:* Dr Shashi, HOD, Surgery, Calicut Medical College, Calicut, India)

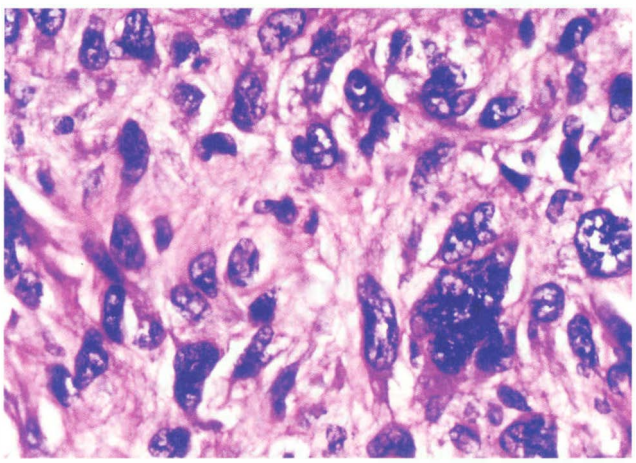


Fig. 15.43: Spindle cell sarcoma (*Courtesy:* Dr Laxmi Rao, Head of the Dept. Pathology, KMC, Manipal)

- Complete tumour resection should be the aim. Chemotherapy and RT are also used.
- Rhabdomyosarcomas have a **high propensity for lymph node metastasis**.

KAPOSI'S SARCOMA

- Vulnerable section of people include Jews, immunocompromised patients such as transplant recipients and AIDS.
- Typical sites: Legs. Other sites include chest, arm, neck in epidemic form (Africa).
- It presents as **multiple pigmented sarcoma nodules** in the leg.
- It is interesting to note that **Kaposi's sarcoma is "not seen" in transfusion related 'AIDS '**.
- It manifests with purplish to red subcutaneous nodules in the leg followed by ulceration and bleeding.
- Combination chemotherapy with doxorubicin, etoposide and interferon have been used to control the disease.

DERMATOFIBROSARCOMA PROTUBERANS
(Fig. 15.39)

- Clinically presents as nodular **exophytic 'mass' lesion—protuberans**.
- Locally aggressive tumour which does not metastasise
- Wide excision should be the aim with negative margin to prevent local recurrence.
- Mohs' micrographic surgery such as basal cell carcinoma has been advocated to get negative margin and thus to get a low recurrence rates.
- Has good prognosis, if treated early.

USEFUL TIPS IN A CASE OF SOFT TISSUE SARCOMA (Key Box 15.13)

In undergraduate clinical examination, students are advised to offer soft tissue sarcoma as the diagnosis. When asked the possible type, they only give a possible histological type based on various clinical features mentioned above. **Ask following questions to yourself** to get ready for the clinical exams.

1. **Is it soft tissue sarcoma?** Tumour arising from soft tissue, dilated veins, reddish skin, increase in local temperature, firm to hard, rapidly growing swelling, with late involvement of skin (carcinoma starts in the skin) (Key Box 15.13).
2. **What is the age of the patient?**
   - In children – Rhabdomyosarcoma
     Undifferentiated sarcoma
   - In 20–40 years – Liposarcoma
     Synovial sarcoma
     Kaposi's sarcoma
   - In elderly patients – Angiosarcoma
     Chondrosarcoma (bone)
     Fibrosarcoma (Fig. 15.23)
3. **Which site has it occurred?**
   - **Head and neck** – Angiosarcoma
     Rhabdomyosarcoma
     Osteogenic sarcoma (jaw)
   - **Distal extremity (limbs)** – Synovial sarcoma (Fig. 15.25)
     Epithelioid sarcoma (Fig. 15.38
     Clear cell sarcoma
   - **Retroperitoneum and Mesentery** – Liposarcoma (Fig. 15.24)
     MFH, leiomyosarcoma
4. **Has it spread to lymph nodes?**
   - Rhabdomyosarcoma
   - Synovial sarcoma
   - Epithelioid sarcoma
5. **Has it spread to lungs or liver?**
   - Chest X-ray
   - Ultrasound
6. **Can I preserve the limb? How?**
   - Wide excision
   - Compartmental excision
   - Preoperative radiotherapy combined with surgery and postoperative radiotherapy.

KEY BOX 15.13

INTERESTING 'MOST' TO REMEMBER IN SOFT TISSUE SARCOMA

- Most of the soft tissue sarcomas **arise *de novo***.
- Most originate in an **extremity** (60%)
- Most common soft tissue sarcoma in adults is **MFH (28%)**.
- Most common soft tissue sarcoma in **children** is **rhabdomyosarcoma**.
- Most of soft tissue sarcomas metastasise by blood.
- Most of the **extremity** soft tissue sarcoma **metastasise to lungs**.
- Most of the **retroperitoneal soft tissue** sarcoma metastasise to liver.
- Most important prognostic factor for soft tissue sarcoma is **size and grade of the primary tumour**.
- Most accurate imaging modality for soft tissue sarcoma is **MRI**.
- Most effective modality of **treatment is surgery**.
- Most effective **chemotherapeutic drugs** are doxorubicin, dacarbazine and ifosfamide.

CLINICAL NOTES

A 34-year-old nondiabetic male patient had a nonhealing ulcer great toe (RT) of 6 months duration. History of minor trauma was present. Examination revealed ulceration, thickening and induration with 'mass' lesion. It was MBBS exam case. The student offered osteomyelitis, foreign body granuloma and tuberculous synovitis as diagnosis. He qualified but he was asked to examine the groin. The patient had 3 hard nodes in the groin. The final diagnosis—Synovial cell sarcoma with metastasis in the groin nodes—the student had not examined the groin!

MULTIPLE CHOICE QUESTIONS

1. When do you suspect lipoma turns into liposarcoma:
A. When lipoma gets infected
B. When lipoma causes lymphangitis
C. When it becomes fixed
D. When it undergoes myxomatous degeneration

2. Intussusception is caused by:
A. Submucosal lipoma
B. Subserosal lipoma
C. Retroperitoneal lipoma
D. Intraperitoneal lipoma

3. Myxomatous degeneration occurs only in which lipoma?
A. Retroperitoneal B. Subfacial
C. Submucosal D. Subcutaneous

4. Following are true for von Recklinghausen's disease *except:*
A. It is an autosomal dominant disorder
B. Skin pigmentation is a feature
C. Café au lait spots are characteristic
D. Does not turn into sarcoma

5. Following are true for Schwannoma *except:*
A. Commonest site is acoustic nerve
B. Sensory branches are affected more often
C. Well encapsulated tumour
D. Highly premalignant tumour

6. Following are radiosensitive tumours *except:*
A. Oral cancer B. Seminoma testis
C. Carcinoma breast D. Malignant chordoma

7. Which one of the following does not have a capsule?
A. Hamartoma B. Schwannoma
C. Fibroadenoma D. Branchial cyst

8. Which one of the following is commonly associated with paraneoplastic syndrome?
A. Carcinoma stomach
B. Carcinoma colon
C. Carcinoma pancreas
D. Carcinoma lung

9. Which one of the following condition is more commonly associated with paraneoplastic syndrome?
A. Hypernatraemia
B. Hyponatraemia
C. Hypercalcaemia
D. Hypocalcaemia

10. Polycythaemia is a paraneoplastic syndrome seen in which condition?
A. Hepatoma B. Wilms' tumour
C. Apudoma D. Renal cell carcinoma

11. Post mastectomy lymphoedema is called:
A. Stewart-Treves syndrome
B. Bloom's syndrome
C. Fanconi's syndrome
D. Sturge-Weber syndrome

12. Precautions to be taken while doing a open biopsy in soft tissue sarcoma include following *except:*
A. Incision should be longitudinal
B. Perfect haemostasis should be achieved
C. Drain should be kept
D. Flaps should not be raised

13. Sarcomas metastasise to lymph nodes include following *except:*
A. Rhabdomyosarcoma
B. Angiosarcoma
C. Synovial sarcoma
D. Liposarcoma

14. Following are true for the treatment of soft tissue sarcomas *except:*
A. Surgery is the best line of treatment
B. Radiotherapy is given after surgery
C. Chemotherapy is also given
D. Radiotherapy is the first line of treatment

15. Following are true for the treatment of retroperitoneal sarcoma *except:*
A. Surgery is the best line of treatment
B. Radiotherapy is extremely helpful
C. Chemotherapy is not very useful
D. Most of them are liposarcoma

16. Following are high grade soft tissue sarcomas *except:*
A. Angiosarcoma
B. Synovial sarcoma
C. Malignant fibrous histiocytoma
D. Liposarcoma

17. Following are true for glomus tumour *except:*
A. It can turn into malignant
B. It is an angioneuromyoma
C. It is radio resistant
D. It is concerned with heat regulation

ANSWERS

1 C	2 A	3 A	4 D	5 D	6 D	7 A	8 D	9 C	10 D
11A	12 C	13 D	14 D	15 B	16 D	17 A			

16

Cystic Swellings, Neck Swellings and Metastasis Lymph Node Neck

- Cystic swellings
- Neck swellings
- Differential diagnosis of midline swellings
- Swellings in the submandibular triangle
- Swellings in the carotid triangle
- Swellings in the posterior triangle
- AV fistula
- Secondaries in the neck
- Different types of neck dissections
- Pancoast's tumour
- What is new?/Recent advances

CYSTIC SWELLINGS

A cyst is a swelling containing fluid. **True cysts** are lined by endothelium or epithelium. They contain clear serous fluid, mucoid material, pus, blood, lymph or toothpaste like material.

The **false cysts** do not have lining epithelium. They can be ***degenerative cysts*** as in the case of tumours which undergo tumour necrosis or tumour degeneration, or merely a ***collection*** of fluid which is walled off by coils of bowel as in tuberculous encysted ascites or an ***exudation cyst*** as in pseudopancreatic cyst.

Classification of cyst

I. Congenital cyst

- Sequestration dermoid cyst
- Branchial cyst
- Thyroglossal cyst
- Lymphangioma
- Cysts of embryonic remnants: Cyst of urachus, vitello-intestinal duct cyst

II. Acquired cyst

- Retention cyst: Sebaceous cyst, galactocoele, spermato-coele, Bartholin's gland cyst
- Distension cyst: Thyroid cyst, ovarian cyst
- Exudation cyst: Hydrocoele
- Degenerative cyst: Tumour necrosis
- Traumatic cyst: Haematoma, implantation dermoid cyst
- Cystic tumours: Cystadenoma of pancreas, cystadenoma of the ovary

III. Parasitic cyst

- Cysticercosis
- Hydatid cyst

Clinical examination of cysts in general

Students are requested to follow the standard practice of examination of the swelling in the form of inspection, palpation, percussion and auscultation in the clinical examination. Some important tests for cystic swellings are given in the next page.

1. **Location:** Most of the congenital cystic swellings have a typical location wherein diagnosis can be made with fair accuracy. A few examples are as follows:
   - **Branchial cyst** occurs at the junction of upper one-third and lower two-thirds of the sternocleidomastoid muscle whereas opening of branchial fistula occurs at junction of upper two-thirds and lower one-third of sterno-cleidomastoid muscle.
   - **Dermoid cyst:** Midline, outer or inner canthus of the eye
   - **Meningocoele:** Swelling in the newborn at lumbosacral region
   - **Ganglion:** On the dorsum of the hand and foot

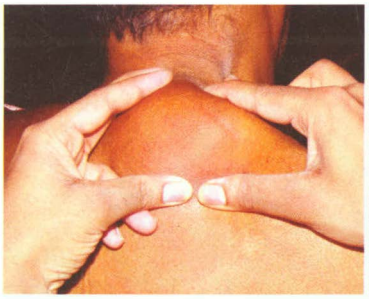


Fig. 16.1: Fluctuation should be elicited using both hands and in both directions

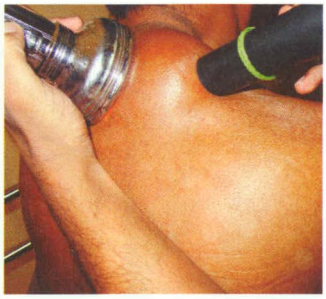


Fig. 16.2: Transillumination test: It should be done in a dark room

2. **Shape:** Majority of the cystic swellings are round or oval
   - **Subhyoid bursitis:** Transverse oval cystic swelling in the midline of the neck.
   - **Thyroglossal cyst:** Vertically placed oval swelling in the midline of the neck.
   - **Sebaceous cyst:** Hemispherical swelling.
3. **Surface:** Almost all the cystic swellings in the skin and subcutaneous tissue have smooth surface.
4. **Consistency:** Fluctuation is positive in all cystic swellings (Fig. 16.1 and Key Box 16.1). However, depending on the contents, the fluctuation may be different, which an experienced surgeon can diagnose.
   - **Soft cystic:** Thyroglossal cyst, meningocoele, lymph cyst.
   - **Tensely cystic:** Ganglion, tensely cystic swellings in the neck may feel firm or solid, e.g. tense thyroid cyst. Cyst in the breast may feel firm or hard.
   - **Yielding** in cases of lipoma, as fat at body temperature behaves like fluid (pseudofluctuation).
   - **Soft with firm thickened periphery:** Cold abscess
   - **Half filled like a rubber hot water bottle:** Branchial cyst
   - **Putty or tooth paste:** Sebaceous cyst (true fluctuation is not found).
   - **Cross fluctuation** for swellings having two components connected to each other, e.g. plunging ranula.
5. **Transillumination test:** Cystic swellings which contain clear fluid show positive transillumination (Fig. 16.2).
6. **Mobility:** Almost all the cystic swellings in the skin, subcutaneous tissue or in the deeper plane are benign and as a rule, they should have free mobility. However, this is not true due to various anatomical factors.
   - **Branchial cyst:** Restricted mobility is due to its adherence to the sternomastoid muscle.
   - **Thyroglossal cyst:** Transverse mobility is absent because the cyst is tethered by remnant of the thyroglossal duct.
   - **Sebaceous cyst:** Limited mobility due to the adherence to the skin.
7. **Sign of compressibility:** The swellings which have communication with a cavity or with tissue spaces give the positive sign of compressibility. Thus, a steady pressure is applied over the swellings. The swelling may disappear completely or may partially disappear. However, when pressure is released the swelling fills up slowly. Hence, it is also called the 'sign of refilling' (Key Box 16.2).
8. **Plane of the swelling**
   - Almost all significant cystic swellings in the neck are deep to deep fascia. Thus, contracting sternomastoid for laterally placed swellings and bending the chin against resistance for centrally placed swellings must be done to define the plane of swelling.
   - Subcutaneous swellings become more prominent when the underlying muscles are contracted as in limbs.
   - Swelling due to semimembranous bursitis, almost disappears on flexion of knee and becomes more prominent on extension of the knee.
   - Sebaceous cysts are attached to the skin at the site of punctum.
9. **Pulsations**
   a. **Expansile:** Aneurysms are characterised by expansile pulsations. When two fingers are placed over the swelling on the sides, the fingers are not only elevated but are also separated. Popliteal aneurysms typically give this sign (Key Box 16.3).
   b. **Transmitted:** When the swelling is situated over a vessel, the fingers are raised but not separated, e.g. pseudopancreatic cysts. When the swelling pushes the vessel anteriorly, transmitted pulsation can be obtained, e.g. cervical rib pushing the subclavian artery.
   c. **Pulsation** can also be present in vascular tumours such as osteogenic sarcoma or secondaries from carcinoma thyroid, etc.

KEY BOX 16.1

RULES OF ELICITATION OF FLUCTUATION

- **Mobile swelling has to be fixed.**
- **Both hands should be used.**
- With the index finger and thumb of one hand the swelling is pressed—these are 'active' fingers and the impulse is received by the thumb and index finger of the other hand (passive fingers).
- Fluctuation should be elicited in **both directions,** as fleshy muscle in the thigh can be fluctuant across but not in the longitudinal direction.
- When swelling is smaller than 2 cm in size, **Paget's test is done**. Cystic swellings feel soft in the centre and firm at the periphery. Solid swellings feel firm at the centre than periphery.

KEY BOX 16.2

COMPRESSIBLE SWELLINGS

- Haemangioma
- Lymphangioma
- Meningocoele

KEY BOX 16.3

ANEURYSM TESTS

- Expansile pulsations: Finger separation sign
- Proximal compression test: Decreased size
- Distal compression test: Size may increase
- Thrill and bruit are present and distal pulses may be weak.

Effects of aneurysm (Key Box 16.4 and Figs 16.3 and 16.4)

KEY BOX 16.4

EFFECTS OF ANEURYSM: TIPS

- Thrombosis, Ischaemia
- Pressure, Skin changes

Some useful TIPS

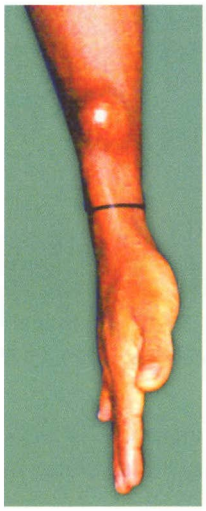
1. **Thrombosis:** It is one of the common effects of aneurysm particularly aortic and popliteal resulting in ischaemia in the distal territory.
2. **Ischaemia:** Distal parts may become gangrenous and or can have ischaemic ulcers or claudication.
3. **Pressure effects**
   a. **Effect on bone:** Erosion of the vertebral body as in aortic aneurysm. This does not happen in TB spine.
   b. **Effect on nerves:** Popliteal aneurysm can give rise to foot drop due to pressure on lateral popliteal nerve.
   c. **Effect on the veins:** Results in congestion and oedema of the leg.
   d. **On the oesophagus:** Dysphagia as in aortic aneurysm.
4. **Skin changes** may be in the form of oedema and redness.

Complications of cysts in general

1. **Infection**, e.g. **sebaceous cyst**
2. **Calcification**, e.g. haematoma, multinodular goitre with cyst, hydatid cyst
3. **Pressure effects:** Ovarian cyst pressing on the iliac veins.
4. **Haemorrhage** within thyroid cyst.
5. **Torsion:** Ovarian dermoid.
6. **Transformation** into malignancy.
7. **Ovarian cachexia:** Large ovarian tumour with pedal oedema, anorexia, loss of weight, lordosis.

DERMOID CYST

This is a cyst lined by squamous epithelium containing desquamated cells. The contents are thick and sometimes toothpaste-like which is a mixture of sweat, sebum and desquamated epithelial cells and sometimes even hair.



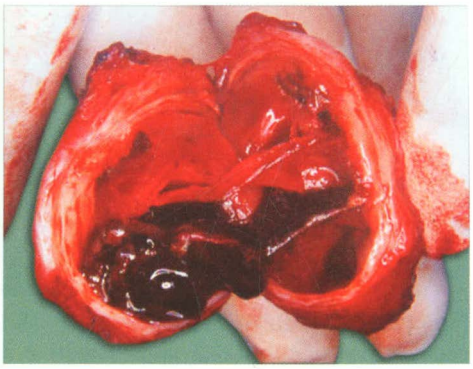


Fig. 16.4: Radial artery aneurysm—with clot—excised specimen (*Courtesy:* Prof L Ramachandra, Professor of Surgery, KMC, Manipal)

Fig. 16.3: Radial artery aneurysm

Clinical types of dermoid cyst (Key Box 16.5)

I. Congenital/sequestration dermoid

- They occur along the line of embryonic fusion, due to dermal cells being buried in deeper plane.
- The cells which are sequestrated in the subcutaneous plane proliferate and liquefy to form a cyst.
- As it grows, it indents the mesoderm (future bone) which explains the bony defects caused by dermoid cyst in the skull or facial bones.
- Even though they are congenital, they manifest as a swelling during childhood or later in life. Often they can be mistaken for lipoma and sebaceous cysts (Fig. 16.5).
- They can occur anywhere in the midline of the body or the face (Key Box 16.5)
  1. External and internal angular dermoid cyst: At the fusion lines of frontonasal and maxillary processes (Fig. 16.6).
  2. Median nasal dermoid cyst: At the root of the nose at the fusion lines of frontal process (Fig. 16.7).
  3. In the suprasternal space of *Burns*
  4. Sublingual dermoid cyst (Fig. 16.8)
  5. Preauricular dermoid cyst—in front of the auricle
  6. Postauricular dermoid cyst behind the auricle (Fig. 16.9).

PEARLS OF WISDOM

Pinna is formed by the fusion of 6 cutaneous tubercles. Both preauricular and postauricular dermoid cysts occur because of failure of fusion of one of the tubercles with the others as they form pinna.

KEY BOX 16.5

ORIGIN OF DERMOID CYST

- The face is developed from 5 processes—2 maxillary, 2 mandibular and 1 frontonasal process.
- Dermoid cyst occurs in the line of embryonic fusion of these processes.

Complications of dermoid cyst

1. Infection
2. Suppuration: Abscess
3. Ovarian dermoid: Torsion

Clinical features

- Though congenital, the cyst manifests in childhood or during adolescence. A few cases also manifest in 30-40 years age group.
- Typically, the patient presents with a painless, slow-growing swelling.
- Soft, cystic and fluctuant; transillumination is negative.
- Rarely, it may be putty like in consistency.
- **The underlying *bony* defect gives the clue to the diagnosis.**
- Classical location of the cyst (along the line of fusion) is a feature of sequestration dermoid cyst.

II. Implantation dermoid cyst (Figs 16.10 and 16.11)

- This is common in women, tailors, agriculturists who sustain repeated minor sharp injuries.
- Following a **sharp injury,** few epidermal cells get implanted into the subcutaneous plane. There, they develop into an implantation dermoid cyst. Hence, it is typically found in the fingers, palm and sole of the foot. As the cyst develops in the areas where the skin is thick and keratinised, it feels firm to hard in consistency.

III. Teratomatous dermoid cyst (Fig. 16.12)

- Teratoma is a tumour arising from ***totipotential cells.*** Thus, it contains ectodermal, endodermal and mesodermal elements—hair, teeth, cartilage, bone, etc.
- Common sites are ovary, testis, retroperitoneum and mediastinum.

Fig. 16.5: Median frontal dermoid cyst—lipoma, sebaceous cyst are other differential diagnosis

Fig. 16.6: External angular dermoid cyst—look for bony depression—it is pathognomonic of this swelling

Fig. 16.7: Median frontal dermoid cyst at the root of the nose. Test for cough impulse

Fig. 16.8: Sublingual dermoid (*Courtesy:* Dr Sreejayan, Professor of Surgery, Calicut Medical College)

Fig. 16.9: Post-auricular dermoid cyst. Location and soft consistency are characteristic

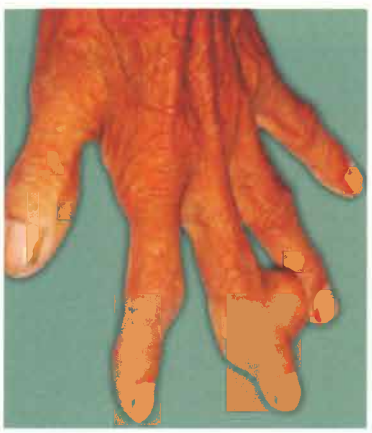


Fig. 16.10: Implantation dermoid cyst—Classical sites are hand and foot which are prone for sharp injuries

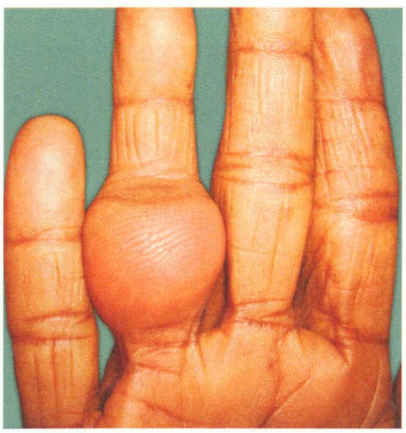


Fig. 16.11: Implantation dermoid cyst—TB synovitis and chronic abscess are the other differential diagnosis

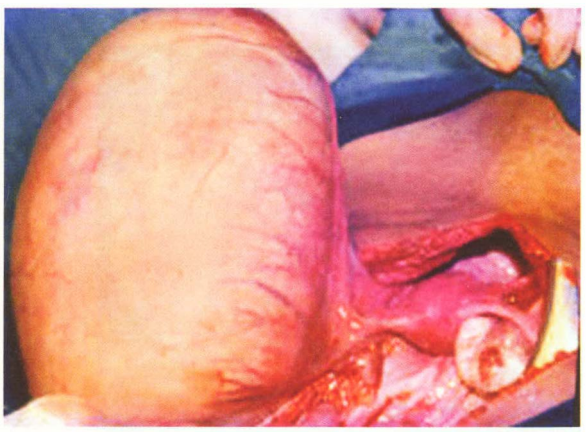


Fig. 16.12: Ovarian teratomatous dermoid cyst. Usually they are bilateral—large ones with pedicles are vulnerable for torsion (*Courtesy:* Dr Rajesh Bhakta, Associate Professor, Dept. of OBG, KMC, Manipal)

CLINICAL NOTES

A 24-year-old female patient was admitted with acute lower abdominal pain of 2 days' duration. There was guarding and rigidity of the abdominal wall with rebound tenderness. Per vaginal examination was normal. At exploration, there was a twisted ovarian teratoma on the left side with gangrene. It was excised. The opposite ovary, on careful examination revealed a small teratomatous dermoid cyst which could be enucleated. The significance of this case report lies in the fact that both ovaries should be examined in cases of ovarian dermoid cysts.

IV. Tubulo-embryonic dermoid cyst

- They arise from ectodermal tubes. A few examples are thyroglossal cyst, post-anal dermoid cyst.
- Ependymal cyst of the brain.

Treatment of dermoid cyst

- Excision of the cyst.

EPIDERMAL CYST (WEN)

- This is popularly called **sebaceous cyst**. It is a misnomer. This occurs due to obstruction to one of the sebaceous ducts, resulting in accumulation of sebaceous material. Hence, this is an example of **retention cyst**.
- **Sites:** Scalp, face, back, scrotum, etc. ***It does not occur in palm and sole***, where sebaceous glands are absent. In the back, scalp and scrotum (Figs 16.15 to 16.19), multiple cysts are often found.

Clinical features

- They are slow-growing and appear in early adulthood or middle age.
- **Hemispherical** or spherical swelling located in the dermis. A dark spot in the centre (punctum) filled with keratin is a diagnostic feature of this cyst. The punctum indicates blockage of the duct (Fig. 16.14).
- In 20–30% of cases, instead of opening into the skin, sebaceous duct opens into the hair follicle. Hence, punctum is not seen.
- It has a smooth surface, round borders, soft and putty consistency and is nontender.
- The cyst can be moulded into different shapes which is described as ***sign of moulding.***
- ***Sign of indentation*** refers to pitting on pressure over the swelling (Table 16.1).
- The swelling is mobile over the deep structures, and the skin is free all around except an area of adherence at the site of punctum.
- In the scalp, loss of hair is a feature over the swelling because of constant slow expansion of the cyst.

Treatment

- Incision and **avulsion of cyst with the wall**. Very often, during dissection, the cyst wall ruptures. Care should be taken to excise the entire cyst wall. If not, recurrence can occur.
- When it is small it can be excised along with the skin.

Complications

1. **Infection** can occur due to injury or scratch resulting in an abscess. The cyst will be tender, red and warm to touch. It should be treated like an abscess by incision and drainage. After one to two months, the cyst can be excised.
2. **Sebaceous horn** results due to slow drying of the contents which are squeezed out, specially if a patient does not wash the part. Thus, it is not common to find a large sebaceous horn nowadays because of better ways of living and sanitation (Fig. 16.20).

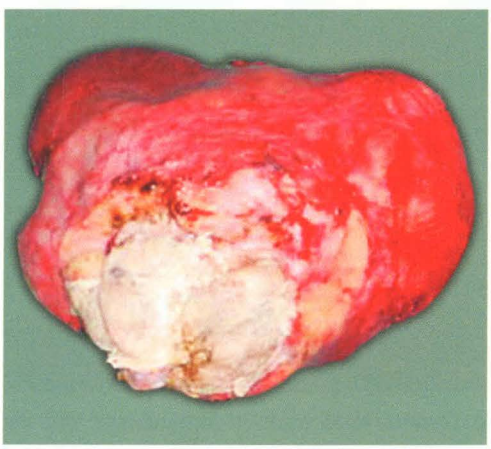


Fig. 16.13: Cut open specimen of epidermoid cyst showing cheesy material

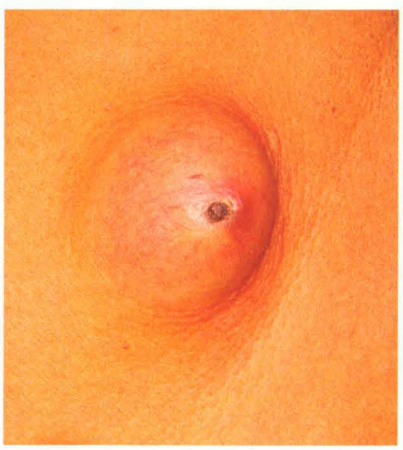


Fig. 16.14: Sebaceous punctum is diagnostic of sebaceous cyst

Fig. 16.15: Multiple sebaceous cysts on the back—troublesome situation

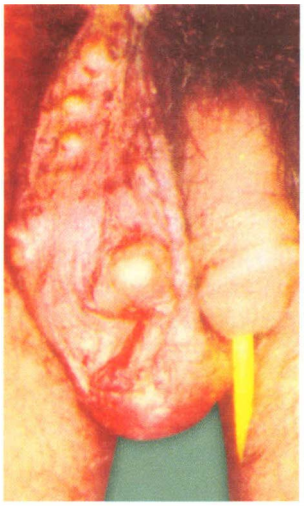


Fig. 16.16: Multiple sebaceous cysts on the scrotum—one of the common sites

Fig. 16.17: Multiple sebaceous cysts on the scrotum—calcified. Requires excision

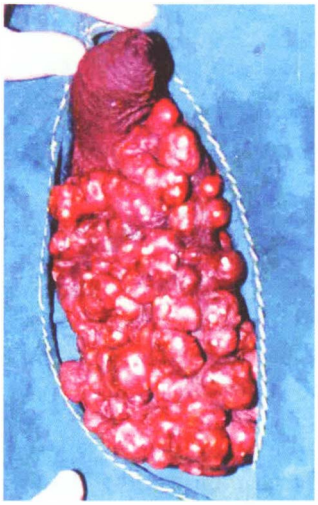


Fig. 16.18: Strawberry scrotum (*Courtesy:* Dr Umesh Bhat, Surgeon, Kundapur, Karnataka, India)

Fig. 16.19: Sebaceous cyst on the scalp—loss of hair is a feature. When punctum is absent, the differentiated diagnosis is lipoma

Table 16.1 Comparison of congenital dermoid cyst and sebaceous cyst

	Congenital dermoid cyst	Epidermal cyst
• Aetiology	Congenital—sequestration of the dermal cells in the subcutaneous plane	Acquired—retention cyst due to accumulation of sebaceous contents
• Location	Midline of the body, along the line of fusion	Face, scalp, scrotum, back
• Sign of indentation, moulding	Uncommon	Very common
• Punctum	Absent	Present in 50% of cases—diagnostic
• Skin fixation	Absent	Skin is fixed at the site of punctum
• Bony defect	Present in majority of cases	Absent
• Intracranial communication	Rare, can be diagnosed by cough impulse test	Absent
• Treatment	Excision	Excision or avulsion

3. Calcification

4. **Cock's peculiar tumour**[1] refers to infected, ulcerated cyst of scalp with pouting granulation tissue and everted edge resembling epithelioma (Key Box 16.6).
5. **Rarely, basal cell carcinoma** can arise in a long-standing sebaceous cyst.

KEY BOX 16.6

INTERESTING—SEBACEOUS CYST

- Syndrome: Gardner's syndrome
- Tumour: Cock's peculiar tumour
- Parasitic worm: Demodex folliculorum
- Strawberry scrotum: Multiple sebaceous cysts of scrotum

Fig. 16.20: Sebaceous horn

[1]Pott's puffy tumour refers to osteomyelitis of the frontal bone with oedema of scalp secondary to frontal sinusitis. This and Cock's peculiar tumour are favourite viva questions.

PEARLS OF WISDOM

Meibomian cyst: They are epidermal cysts found on the free edge of eyelid. Chronic Meibomian cyst is called chalazion.

GANGLION

It is a tense, cystic swelling and occurs due to myxomatous degeneration of the synovial sheath lining the joint or tendon sheath. They are common around joints because of abundant fibrous tissue. They contain gelatinous fluid.

Common sites

- The dorsum of the hand is the common site, at the **scapholunate** articulation.
- In the foot, dorsal or lateral aspect.
- Small ganglion in relation to flexor aspect of fingers.

Clinical features

- Majority of patients are between 20 and 50 years.
- A round to oval swelling in the dorsum of the hand, with smooth surface and round borders. Skin over the swelling is normal.
- The swelling is tensely cystic and fluctuant. Trans-illumination is negative. It is mobile in the transverse direction.
- When the tendons are put into contraction, the mobility of the swelling gets restricted.
- Ganglion is not connected with the joint space. Sometimes, it gives an impression of becoming small due to slipping away between bones.

Treatment

1. Asymptomatic ganglion is ***better left alone***.
2. ***Aspiration*** of the ganglion and injection of sclerosants may reduce the size of ganglion.
3. Sometimes, rupture of the cyst due to trauma may result in permanent cure.
4. ***Surgical excision*** can be done. However, recurrence rate is high.

Differential diagnosis (DD)

1. Implantation dermoid cyst, when it occurs in the feet or hand.
2. Exostosis of the bone, has to be considered if *swelling is very hard.*
3. Bursa (*vide infra*)

COMPOUND PALMAR GANGLION

Aetiology

- ***Tuberculous tenosynovitis*** of the tendon sheaths affecting the flexor tendons. This is a common cause in India (Key Box 16.7)
- ***Rheumatoid arthritis*** with involvement of multiple joints causing thickening of synovial membrane—common cause in Western countries.

Pathology

- As a result of tuberculous tenosynovitis, typical caseous material collects within the flexor tendon sheaths. The tendons get matted, a swelling develops in the palm and another swelling develops in lower aspect of forearm. The thickening of synovial membrane, fibrin particles in the **fluid and melon seeds**, are characteristic of this condition.

Clinical features

- Majority of patients are below 40 years of age.
- Concavity of the palm is obliterated.
- Soft, cystic, fluctuant, transillumination—negative swelling situated above and below the flexor retinaculum.
- **Cross fluctuation test** between these two swellings is positive, which is diagnostic of compound palmar ganglion.
- Restricted mobility of the fingers due to matting of the tendons.
- **Wasting of the small muscles** of the hand.
- **Paraesthesia** due to compression on median nerve.

Investigations

1. The ESR may be increased if it is due to tuberculosis.
2. Aspiration of the swelling and fluid can be sent for acid-fast bacilli.
3. Synovial biopsy.

Treatment

1. **Antituberculous treatment (ATT)** in case of tubercular pathology. If the response rate is not satisfactory—exploration, decompression, synovectomy and release of matted tendons is the treatment.
2. **Control of rheumatoid arthritis**, with complete excision of the synovial sheath, in cases due to rheumatoid arthritis.

Summary of compound palmar ganglion

- Tuberculosis and rheumatoid arthritis—common causes.
- Synovial thickening will clinch the diagnosis.
- Cross fluctuation test is an important clinical finding.
- Antituberculous treatment if it is due to tuberculosis.
- Decompression or synovectomy may be required in both conditions mentioned above.

GLOMUS TUMOUR (Key Box 16.7)

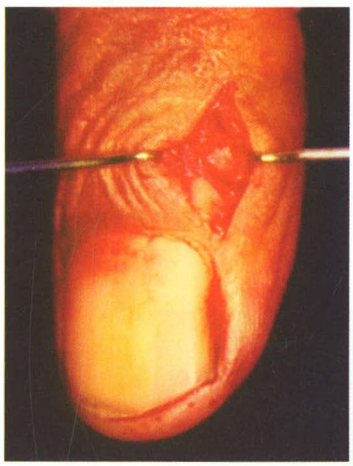
- This is also called glomangioma or angioneuromyoma.
- Glomus is a specialised organ.

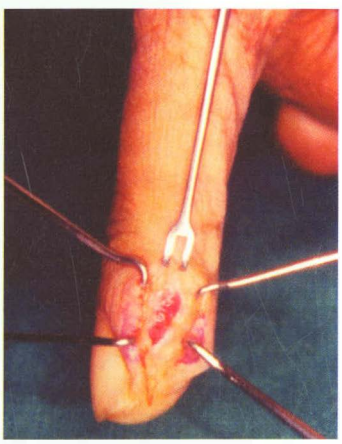
Structure of glomus (glomus body)

Abundant arteriovenous anastomosis surrounded by large clear cells (glomus cells) and medullated and non-medullated nerve fibres in between the cells is characteristic of glomus.

Clinical features of glomus tumour
(Figs 16.21 and 16.22)

1. Typical site: Under the nail beds of hands and feet.
2. It is purple red in colour, usually single, the size does not exceed 1 cm in diameter.
3. Glomus tumour is usually seen in the 5th decade.
4. Excruciating pain either at rest or on movement of the finger or on pressure is pathognomonic feature of this tumour. Pain is due to compression of the nerve fibres by dilated glomus vessels.
5. The tumour is compressible.





Figs 16.21 and 16.22: Glomus tumour: The most painful condition in the finger (*Courtesy:* Prof Bhaskarananda Kumar and Dr Anil Bhat, Department of Orthopaedics, KMC, Manipal)

KEY BOX 16.7

GLOMUS TUMOUR

- Rare and benign tumour
- The **most painful** tumour
- The **smallest benign tumour,** does not turn malignant
- **Nail bed** is the commonest site
- Histologically, it is an **angioneuromyoma**
- It is **radioresistant**
- **Excision** gives permanent cure
- Function of glomus is concerned with **heat regulation.**

Treatment

- **Surgical excision** results in *permanent cure.*

Differential diagnosis

1. Subungual melanoma: Painless and pigmented
2. Granuloma pyogenicum: Mild pain, bleeds on touch and evidence of infection is present.
3. Chronic infection with granuloma.

BURSA

- Bursa means a sac or a sac-like cavity containing fluid lined by endothelium. It is meant to reduce the friction between tendons of the muscle and the bone.
- Bursitis refers to inflammation of a bursa resulting in accumulation of excessive fluid inside the bursa. This results in a swelling in the anatomical sites of normal bursa.
- The causes of chronic bursitis includes constant pressure, constant irritation or minor injuries.
- Some examples of bursitis are given in Table 16.2.

Clinical features

- A cystic swelling in a known anatomical site of a bursa is a chronic bursitis unless proved otherwise.

Table 16.2 Bursae and bursitis

Anatomical site	Popular nomenclature
1. Prepatellar bursa	Housemaid's knee
2. In front of patella tendon (infrapatellar)	Clergyman's knee
3. Olecranon bursa	Student's elbow
4. Under the insertion of tendons of sartorius, gracilis and semitendinosus muscle	Bursa anserina (extension of the bursa along the sides of tendon-resembles goose's foot)
5. Between the tendon of the semimembranosus and the medial condyle of tibia	Semimembranosus bursitis

- Bursitis produces a soft, cystic, circumscribed or oval swelling with fluctuation.
- As majority of bursitis contains inflammatory fluid, they do not show transillumination.
- In a few cases, signs of inflammation may be present.

Complications

1. Secondary infection may result in an abscess
2. Frequent friction may result in ulceration
3. Cosmetic deformity.

Treatment

- Excision is indicated only in the presence of symptoms such as pain or complications mentioned above.
- Chances of recurrence are high.

SEMIMEMBRANOSUS BURSA (Figs 16.23 to 16.25)

This is the commonest swelling in the popliteal space. It presents as a tensely cystic swelling when the knee is extended and it becomes flaccid on flexion of the knee. It is not compressible as it does not communicate with the joint.

The differential diagnosis for semimembranosus bursitis is **Morrant-Baker's cyst**, which is a herniation of the synovial membrane. The differences between these two swellings are given in Table 16.3.

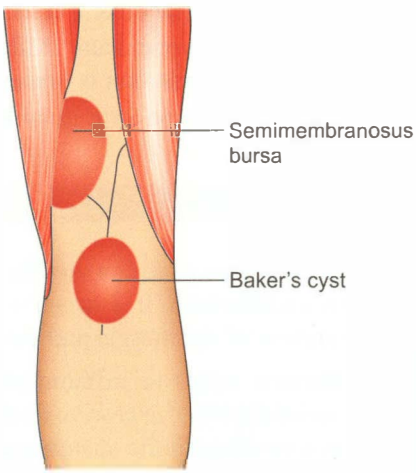


Fig. 16.23: Popliteal fossa swelling

Adventitious bursae

- This refers to a cyst which develops in an anatomical area where no bursa is present. These also occur due to constant pressure or friction. They are summarised below.

1. Tailor's ankle: Above the lateral malleolus

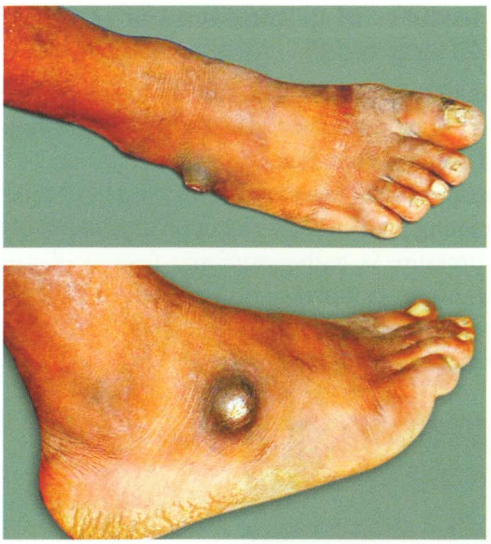
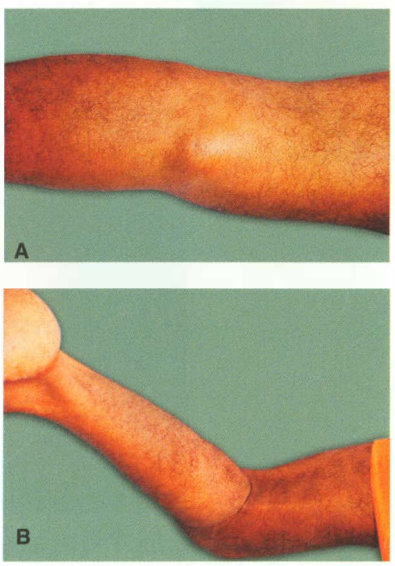


Fig. 16.24: Constant pressure on the lateral aspect of foot resulting in bursa due to the habit of 'squatting position'. Second picture showing the bursa with pigmentation all around



Figs 16.25A and B: (A) Semimembranosus cyst and (B) cyst disappears on flexing

Table 16.3 Comparison of semimembranosus bursa and Baker's cyst (Fig. 16.23)

	Semimembranosus bursa	Baker's cyst
1. Aetiology	Friction or pressure	Rheumatoid or osteoarthrosis of knee joint
2. Age	Young patients	Middle aged
3. Location in the popliteal fossa	Higher up and more medial	Below and midline
4. On flexion of the knee	Disappears	Increases
5. On extension of the knee	Appears and is tense (Figs 16.25A and B)	Diminishes
6. Patellar tap	Absent	Present
7. Compressibility	Absent	Present partially
8. Knee movements	Normal	Restricted

2. **Porter's shoulder:** Between clavicle and skin
3. **Weaver's bottom:** Between gluteus maximus and ischial tuberosity
4. **Bunion:** Between prominent head of the first metatarsal and skin due to hallux valgus

- The complications and treatment of adventitious bursae are similar to chronic bursitis (Fig. 16.24)

TRANSILLUMINANT SWELLINGS IN THE BODY

These are the cystic swellings containing clear fluid characterised by **fluctuation and transillumination**.

1. Lymphangioma
2. Ranula
3. Meningocoele
4. Epididymal cyst
5. Vaginal hydrocoele.

LYMPHANGIOMA (Figs 16.26 to 16.29)

- Failure of one of the lymphatics to join the major lymph sac of the body results in a lymphangioma. Hence, it occurs in places where lymphatics are abundant.
- They are dilated lymphatics that project onto the skin surface.
- Common sites: Posterior triangle of the neck, axilla, mediastinum, groin, etc. (Key Box 16.8).
- In the neck, it is called cystic hygroma of the neck. As the sac has no communication with lymphatics by the time swelling appears, the lymph is absorbed and is replaced by thin watery fluid (mucus) secreted by endothelium. Hence, it is also called hydrocoele of the neck.
- When it is largely confined to subcutaneous plane, it is called **cystic hygroma**.

KEY BOX 16.8

LYMPHANGIOMA	SITES
• Jugular lymph sac	Neck
• Posterior lymph sac	Groin
• Cisterna chyli	Retroperitoneum

Types of lymphangioma

1. **Lymphangioma circumscriptum:** If it is less than 5 cm across.
2. **Lymphangioma diffusum:** If they are more widespread.
3. **Lymphoedema *ab igne*:** If they form a reticulate pattern of ridges.

Clinical features

- Usually, cystic hygroma presents during infancy or early childhood. Occasionally, present since birth and rarely before birth. They can also present as small vesicles.
- When the child cries or strains the swelling increases in size and becomes prominent due to increased intra-thoracic pressure which is transmitted through root of the neck.
- Typical locations—lateral aspect of neck (posterior triangle), groin, buttocks.
- Soft, cystic, fluctuant, partially compressible swelling. Lymphangioma is a multilocular swelling consisting of aggregation of multiple cysts. These cysts may inter-communicate and may occasionally insinuate between muscle planes. Hence, it gives the sign of compressibility! However, **complete reducibility is not a feature**.
- The swelling is **brilliantly transilluminant** because it contains clear fluid (watery lymph) (Key Box 16.9).

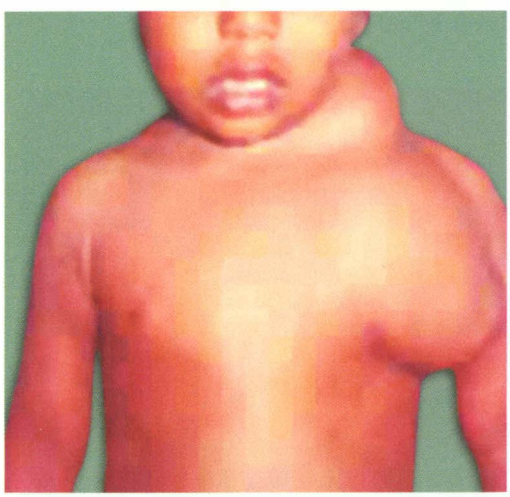


Fig. 16.26: Lymphangioma involving chest wall, neck. The two were different swellings

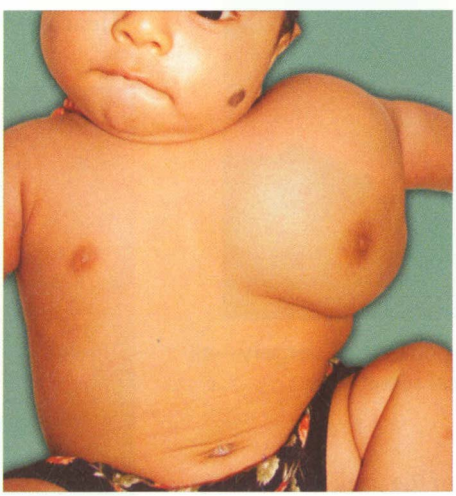


Fig. 16.27: Lymphangioma involving chest wall. Brilliantly transilluminant

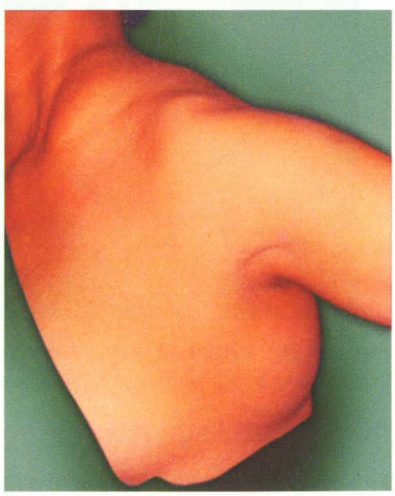


Fig. 16.28: Lymphangioma involving neck and axilla—cross fluctuation was positive

KEY BOX 16.9

TRANSILLUMINATION TEST

- Should be done in a dark room
- Avoid surface transillumination
- Transillumination may be negative because of infection, sclerotherapy and haemorrhage

Treatment

- **Surgical excision** is the treatment of choice. **All the loculi or cysts should be removed.** Careful search has to be made for the extension of lymphangioma through the **muscle planes** so as to avoid **recurrence** (Figs 16.29 and 16.30). Sclerotherapy was being used earlier for lymphangioma. Since, tissue planes are distorted by sclerosants, dissection becomes difficult. **Thus, injection type of treatment is not favoured at present**.

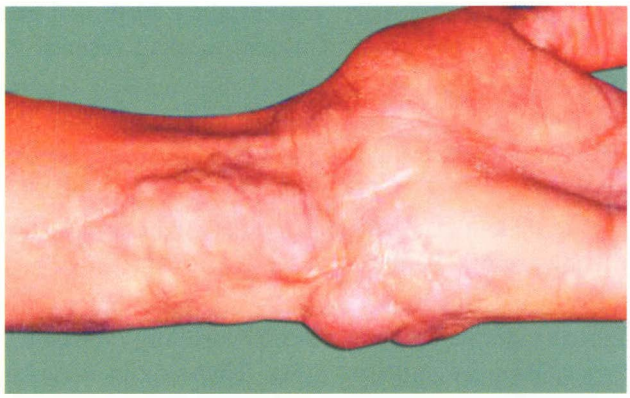


Fig. 16.29: Lymphangioma twice excised—residual lesion. It can be left alone if it is asymptomatic

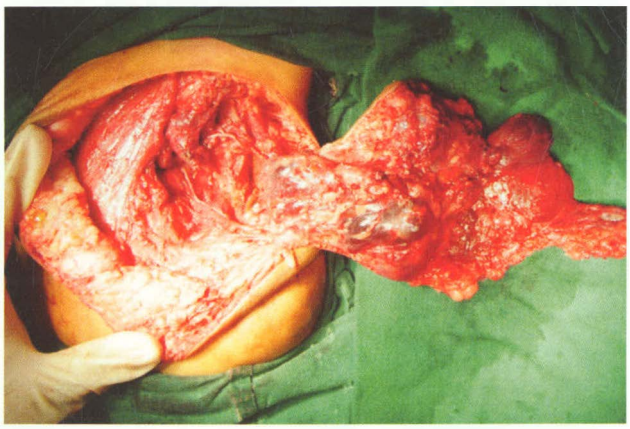


Fig. 16.30: Lymphangioma—multiple pockets at surgery—all those have to be removed to prevent local recurrence

Differential diagnosis

1. **Haemangioma:** Posterior triangle of the neck is one of the common sites for haemangioma. Haemangioma is soft, cystic and fluctuant but transillumination is negative and the sign of compressibility is positive.
2. **Lipoma:** This is a soft lobular swelling with fluctuation because fat behaves like fluid at body temperature. However, the edge slips under the palpating fingers. Both transillumination and compressibility tests are negative with lipoma.
3. **Cold abscess** (details follow in a later page).

Complications

1. In neonates and infants, lymphangioma can cause difficulty in breathing due to its large size.
2. Occasionally, secondary infection can occur.
3. Lymphangioma in the mediastinum can give rise to dyspnoea, dysphagia due to compression on the trachea/oesophagus.

RANULA

Ranula is a cystic swelling arising from sublingual salivary gland and from accessory salivary glands which are present in the floor of the mouth called **glands of Blandin and Nuhn**.

The word ranula is derived from the resemblance of the swelling to the **belly of frog—*Rana hexadactyla***.

Aetiology

1. Ranula occurs due to obstruction to the ducts secreting mucus. Hence, it is an example for ***retention cyst.***
2. Some surgeons consider it as an ***extravasation cyst.***

Clinical features

- Seen in young children and adults.
- The swelling is typically located in the floor of the mouth or **under surface of the tongue,** to one side of the midline (Fig. 16.31).
- Soft, cystic, fluctuant swelling, which gives **brilliant transillumination.**
- It is covered by thin mucosa containing clear, serous fluid. Hence, it is **bluish in colour** (Fig. 16.32).

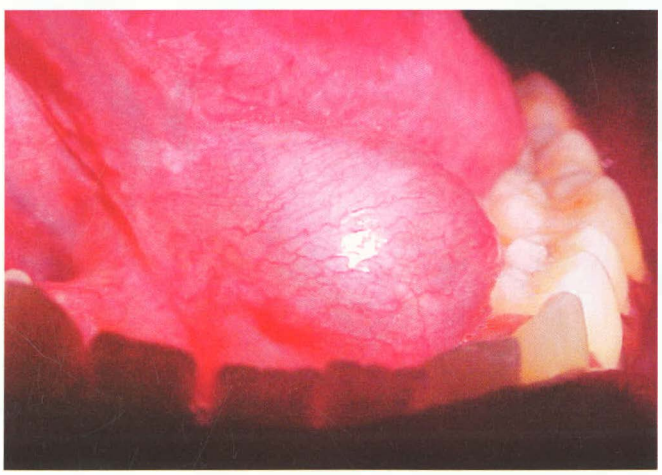


Fig. 16.31: Ranula in the floor of the mouth

Fig. 16.32: Ranula—observe the bluish colour

- Surface is smooth, borders are diffuse, nontender swelling.
- ***Plunging ranula:*** It is an intraoral ranula with cervical extension, where it passes on the side of mylohyoid muscle and produces a swelling in the submandibular region. Thus, one swelling in the floor of the mouth and the other in the neck gives rise to plunging ranula. The diagnosis is confirmed by cross-fluctuation test (Key Box 16.10).

KEY BOX 16.10

CROSS-FLUCTUATION TEST

- Indicated when a cyst has two interconnected components
- Gentle pressure on one component, impulse felt on the other component
- Demonstrated by bidigital palpation
- Plunging ranula, compound palmar ganglion, iliopsoas abscess, hydrocoele *en bisac* will give a cross-fluctuation test.

Treatment

1. **Complete excision** of the ranula is the treatment of choice in plunging ranula. Since the cyst wall is very thin, it should be carefully dissected and removed.
2. **Marsupialisation** is indicated in simple ranula. The ranula is incised and the wall of the cyst is sutured to the mucosa of the floor of the mouth, so as to leave an opening to the exterior (Marsupials, e.g. kangaroo).
   - After 5–10 days, the cyst gets collapsed, fibrosis occurs and the entire cavity gets obliterated.
   - **Marsupialisation avoids surgical dissection** and chances of injury to the submandibular duct.
   - **Plunging ranula can be excised by intraoral approach.** Once the intraoral dissection is completed the cervical extension can be mobilised by the same incision dissecting close to the cyst wall. However, rupture and chances of leaving behind a portion of the cyst wall are high.

Differential diagnosis

- Sublingual dermoid cyst is a thick-walled cyst, whitish in colour and **not transilluminant**.
- Mucus cyst

Complications

1. Rupture of the cyst decreases the size but it can reappear at a later date.
2. When the swelling is big, the tongue is pushed upwards and may cause difficulty in speech or swallowing.

MENINGOCOELE

Meningocoele is a herniation of the meninges through a weak point in the spine (neural arch) where the bony fusion has not taken place effectively (Fig. 16.33 and Key Box 16.11). The swelling is covered by pia mater and arachnoid mater without a dural covering. The swelling contains cerebrospinal fluid (CSF). **Meningocoele is an example of spina bifida cystica.**

Clinical features

- The swelling is present **since birth**.
- Soft, cystic, **fluctuant with brilliant transillumination** are the typical features of the swelling.
- Sign of **compressibility** is present due to displacement of CSF.
- When the child cries or coughs, an **expansile impulse** is present.
- On palpating the edge of the swelling, a bony defect is usually found.

KEY BOX 16.11

MENINGOCOELE: SITES

- Lumbosacral: The commonest
- Occipitocervical: Second common
- Root of the nose: Rare

Fig. 16.33: Meningocoele

Fig. 16.34: Encephalocoele (*Courtesy:* Prof Vijaykumar, Paediatric Surgeon, KMC, Manipal)

Treatment (Key Box 16.12)

- CT scan is done to look for hydrocephalus. If it is present, a ventriculoperitoneal shunt is done which will reduce the meningocoele.
- Excision of the meningocoele should be done as early as possible to prevent the rupture and secondary infection.

Complications

- Skin covering the swelling is very thin and so is prone to ulceration. Due to ulceration, secondary infection and meningoencephalitis can occur.
- Haemorrhage.

KEY BOX 16.12

EXCISION OF MENINGOCOELE

- Surgery: As early as possible after birth
- Early closure prevents infection
- Transverse elliptical incision
- Excision of the sac
- Closure of the defect by plication
- Approximation of the muscles

SPINA BIFIDA OCCULTA

- In this condition, the neural arch is defective posteriorly. There is no visible swelling.
- It can be suspected when there is a tuft of hair, lipoma, naevus, pigmented patch of skin overlying the lumbosacral region.
- Child is normal at birth. Neurological symptoms such as weakness, sciatica-like pain may start appearing at puberty (neurogenic talipes equinus—club foot).
- During this time because of growth there may be traction on the spinal cord by a ligament called *membrane reuniens*.
- X-ray can demonstrate the bifid spine.
- Surgical excision of the membrane gives permanent cure to the patient, if there are symptoms.

Types of spina bifida cystica

1. **Menigocoele**
2. **Meningomyelocoele**

} *See* Table 16.4 for comparison.

- Protrusion of meninges, with ***nerve root of*** **spinal cord or disordered spinal cord** results in meningomyelocoele.
- Neurological deficit such as **foot drop, talipes, trophic ulcer** of the foot (S_1 root) may be present.
- Surgical excision may be followed by residual neurological deficit.

3. **Syringomeningomyelocoele**
   - In this condition, in addition to the meninges, the central canal of spinal cord is also herniated out.
   - Most of the children are stillborn.
   - Very difficult to treat, if the child survives.

Encephalocoele (Fig. 16.34): It is also known as cranium bifidum. It is a neural tube defect characterised by sac-like protrusion of brain and meninges through an opening in the skull.

DIFFERENTIAL DIAGNOSIS OF MIDLINE SWELLING IN THE NECK (*see* page 246)

Midline swellings: From above downwards

1. Ludwig's angina
2. Enlarged submental lymph nodes

Table 16.4 Comparison of meningocoele and menigomyelocoele

	Meningocoele	Meningomyelocoele
Contents	Membranes	Membranes with nerve roots
Consistency	Soft and cystic	Soft to firm
Transillumination	Brilliant	Partially transilluminant
Longitudinal furrow	Absent	Present due to adherence of the nerve roots to the skin
Neurological deficit	Absent	Trophic ulcers, bladder and bowel incontinence, locomotor problems are present
Prognosis after repair	Good	Residual neurological deficit is present

3. Sublingual dermoid cyst
4. Subhyoid bursitis
5. Thyroglossal cyst
6. Enlarged isthmus of thyroid gland
7. Pretracheal and prelaryngeal lymph nodes
8. Retrosternal goitre
9. Thymic swelling
10. Swelling in the suprasternal space of Burns: Lipoma/cold abscess/aneurysm.

LUDWIG'S ANGINA

- This is an inflammatory oedema of the floor of the mouth. It spreads to the submandibular region and submental region.
- Tense, tender, brawny, oedematous swelling in the submental region with putrid halitosis is characteristic of this condition.

ENLARGED SUBMENTAL LYMPH NODES

The three important causes of enlargement:

1. **Tuberculosis: Matted submental nodes,** firm in consistency, with enlarged upper deep cervical lymph nodes, with or without evening rise of temperature are suggestive of tuberculosis.
2. **Non-Hodgkin's lymphoma** can present with submental nodes along with other lymph nodes in the horizontal group of nodes such as submandibular, upper deep cervical, pre-auricular, post-auricular and occipital lymph nodes (external Waldeyer's ring). Nodes are ***firm or rubbery,*** **discrete without matting.**
3. **Secondaries** in the submental lymph nodes can arise from carcinoma of the ***tip of the tongue, floor of the mouth, central portion of the lower lip.*** The nodes are **hard** in consistency and sometimes, **fixed.**

SUBLINGUAL DERMOID CYST

- It is a type of **sequestration dermoid cyst** which occurs due to sequestration of the surface-ectoderm at the site of fusion of the two mandibular arches. Hence, such a cyst occurs in the midline, in the floor of the mouth (Key Box 16.13).
- When they arise from **2nd branchial cleft,** they are found lateral to the midline. Hence, **lateral variety**.
- The cyst is lined by squamous epithelium and contains hair follicles, sebaceous glands and sweat glands. **It does not contain hair**.

Clinical features

1. Young children or patients between the age of 10 and 20 years present with painless swelling in the **floor of mouth**.

KEY BOX 16.13

SUBLINGUAL DERMOID CYST

- Origin: At the site of fusion of 2nd branchial arches
- Site: **Midline**—common; **Lateral**—uncommon
- **Supraomohyoid** variety is common
- **Bidigital palpation** for demonstration of fluctuation
- Soft, cystic, fluctuant, transillumination negative swelling

Differential diagnosis

- Ranula: Transillumination is positive
- Thyroglossal cyst: Moves with deglutition

2. Swelling is **soft** and **cystic**. **Fluctuation** test is positive. **Bidigital palpation** gives a better idea about fluctuation with one finger over the swelling in the oral cavity and the other finger in the submental region.
3. **Transillumination test is negative** as it contains thick, cheesy, sebaceous material.

Differential diagnosis

1. **Ranula:** When a sublingual dermoid cyst is in the midline in the floor of the mouth and above the mylohyoid muscle, ranula is considered as differential diagnosis. However, **ranula is bluish in colour, brilliantly transilluminant.**
2. **Thyroglossal cyst** should be considered as differential diagnosis when the sublingual dermoid cyst is below the mylohyoid muscle. Thyroglossal cyst **moves up with deglutition** whereas **a sublingual dermoid cyst does not.**

Treatment

- Through intraoral approach, excision can be done for both types of sublingual dermoid cyst.

SUBHYOID BURSITIS

- Accumulation of inflammatory fluid in the subhyoid bursa results in a swelling and is described as subhyoid bursitis.
- The bursa is located below the hyoid bone and in front of thyrohyoid membrane.

Clinical features

- The swelling is in front of the neck, in the midline below the hyoid bone (Fig. 16.43).
- The ***swelling is oval in the transverse direction.***
- It moves up with deglutition.
- Soft, cystic, fluctuant and transillumination negative swelling ***(turbid fluid).***
- The swelling ***may be tender*** as it contains inflammatory fluid.

A FEW MIDLINE SWELLINGS IN THE NECK (Figs 16.35 to 16.42)

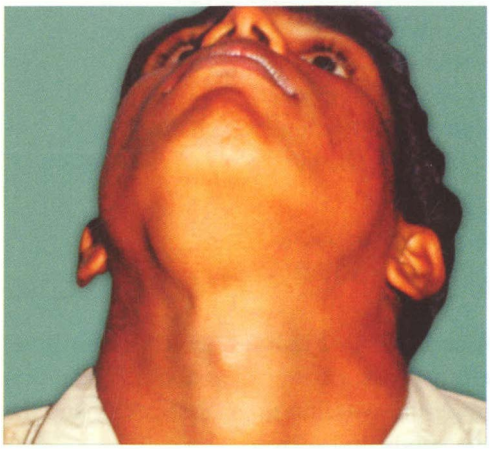


Fig. 16.35: Submental lymphadenitis: Caused by tuberculosis—not a common site

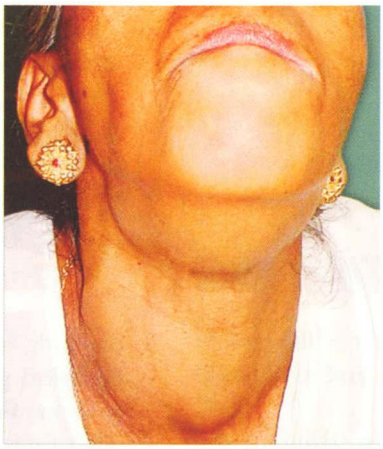


Fig. 16.36: Submental swelling of 30 years duration (*Courtesy:* Prof. KK Rajan, Calicut Medical College)

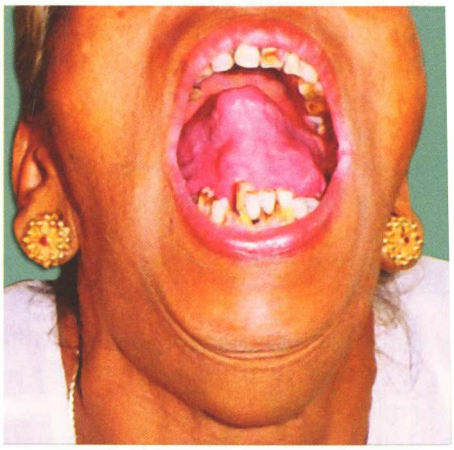


Fig. 16.37: Intraoral examination of patient (Fig. 16.36) reveals swelling also in the floor of mouth—sublingual dermoid cyst

Fig. 16.38: Incompletely treated thyroglossal cyst. Recurred, about to rupture and form thyroglossal fistula

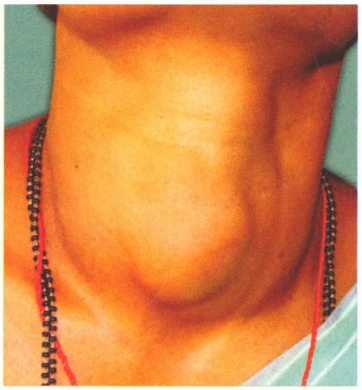


Fig. 16.39: Thyroid nodule: common swelling—moves with deglutition

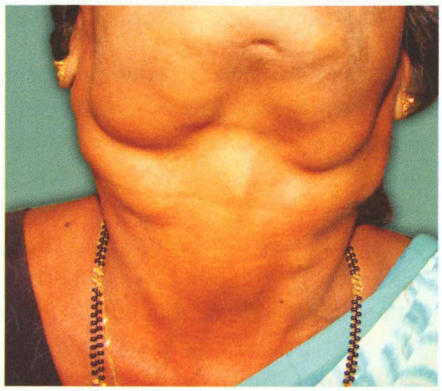
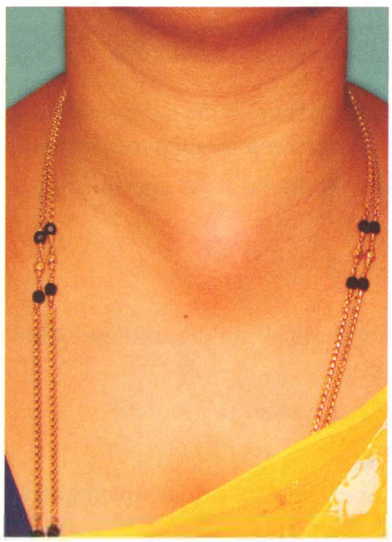
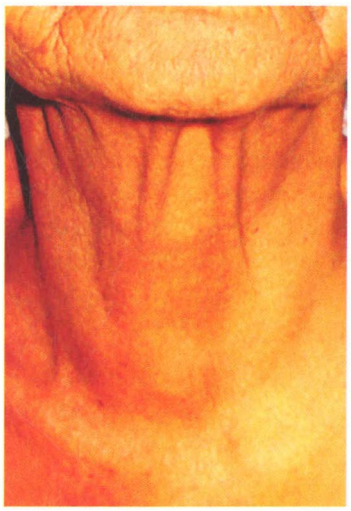


Fig. 16.40: Pretracheal lymph nodes and upper neck nodes: Case of non-Hodgkin's lymphoma



2008: MS Examination
short case, KMC, Manipal (*Courtesy:* Professor BH Ananda Rao, Department of Surgery)

Fig. 16.41: Cold abscess in the suprasternal space of burns: One of the common sites of cold abscess



2008: MS Examination
short case, Govt. Medical College, Goa (*Courtesy:* Prof Dilip Amonkar, HOD, Dept. of Surgery)

Fig. 16.42: Aneurysm of innominate artery. Atherosclerosis is the cause. Before incising a cold abscess in this location, make sure that it is not an aneurysm

Treatment

Complete excision

Complication

It can develop into an abscess

Differential diagnosis

1. Thyroglossal cyst is a vertically placed oval swelling, whereas **subhyoid bursitis** is transversely placed oval swelling (Fig. 16.44).
   - Thyroglossal cyst moves on protrusion of the tongue outside (subhyoid bursitis does not).
2. Pretracheal lymph node swelling.
3. Ectopic thyroid enlargement.

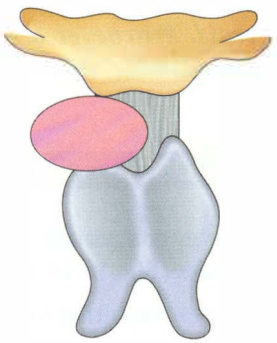


Fig. 16.43: Subhyoid bursitis: Transversely placed oval swelling

THYROGLOSSAL CYST

- This is an example for **tubulo-embryonic dermoid cyst.**
- It arises from *thyroglossal tract/duct* which extends from **foramen caecum at the base of the tongue to the isthmus of the thyroid gland.** Hence, the thyro-glossal cyst can develop anywhere along this duct. For anomalies are shown in Fig. 16.45.
- It is lined by **pseudostrati-fied, ciliated, columnar** or squamous epithelium which produces desquamated epithelial cells or mucus at times.

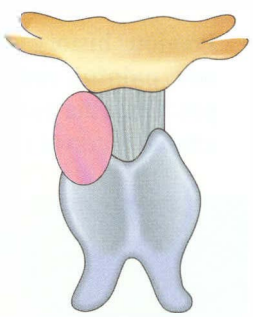


Fig. 16.44: Thyroglossal cyst: Vertically placed oval swelling

Sites of thyroglossal cyst (Fig. 16.46)

1. Subhyoid: **The most common type**
2. At the level of thyroid cartilage: 2nd common site
3. Suprahyoid: Double chin appearance
4. At the foramen caecum: Rare
5. At the level of cricoid cartilage: Rare
6. In the floor of the mouth

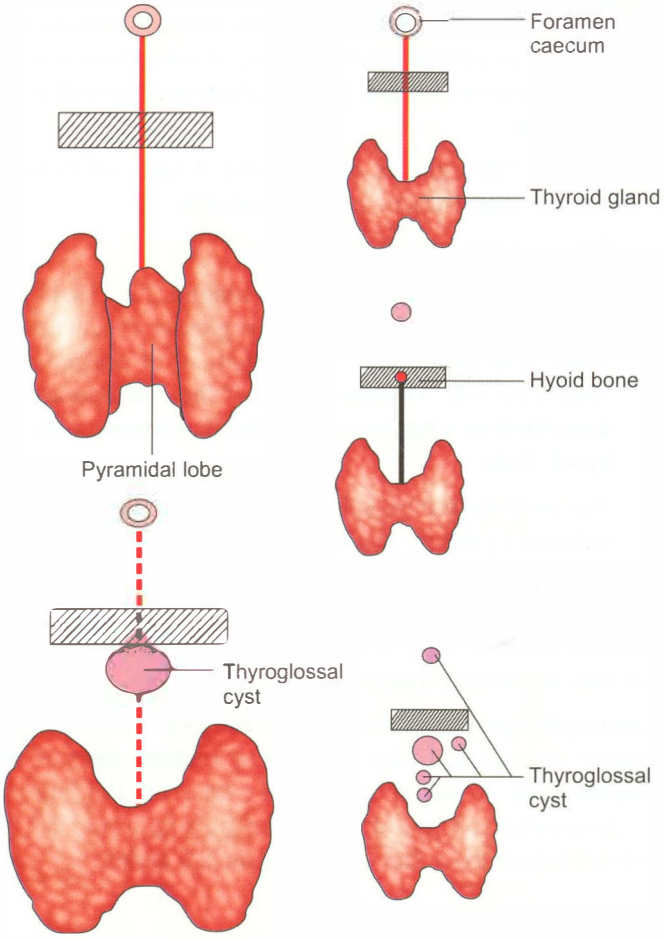


Fig. 16.45: Anomalies/fate of thyroglossal tract

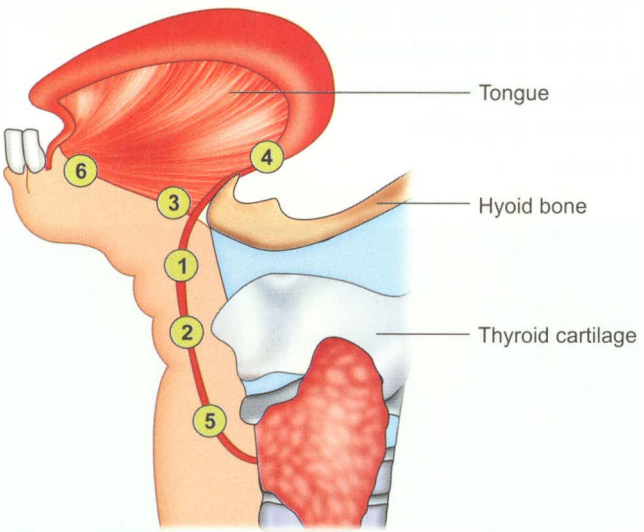


Fig. 16.46: Sites of thyroglossal cyst (numbers *see* text)

Clinical features

- Even though congenital, thyroglossal cyst appears around the age of 15–30 years.
- They are more common in females who present with painless, midline swelling. However, in the region of thyroid cartilage, the swelling is slightly deviated to the left side.

- The cyst is soft, cystic, fluctuant, transillumination—negative swelling (very rarely, it can give rise to transillumination). It can be firm if the tension within the cyst is high.
- **Mobility:** Thyroglossal cysts exhibit 3 types of mobility which are characteristic of this condition:
  - The cyst moves with deglutition.
  - **Moves with protrusion of the tongue** (Fig. 16.47 and Key Box 16.14): Hold the thyroglossal cyst with the finger and thumb and ask the patient to protrude the tongue outside. **The movement of the cyst upwards is described as a tug because of its attachment with the hyoid bone**.
  - The swelling moves sideways but not vertically as it is tethered by the thyroglossal duct.

Treatment

- Before excision of cyst, a thyroid scan is mandatory since it may be the only functioning thyroid tissue.
- **Sistrunk operation: Excision of the cyst** along with the entire thyroglossal tract which may **include part of the hyoid bone**, is the recommended treatment. The intimate relationship of hyoid bone can be explained by its development from 2nd and 3rd branchial arches.

Complication

1. **Recurrent infection:** The wall of the thyroglossal cyst sometimes contains **lymphoid tissue** which can get infected, resulting in an **abscess**. If it ruptures or is incised it results in **thyroglossal fistula** (Key Box 16.15).
2. Rarely, a **papillary carcinoma** can occur in the **thyroglossal cyst** (*see* page 313).
3. Fistula.

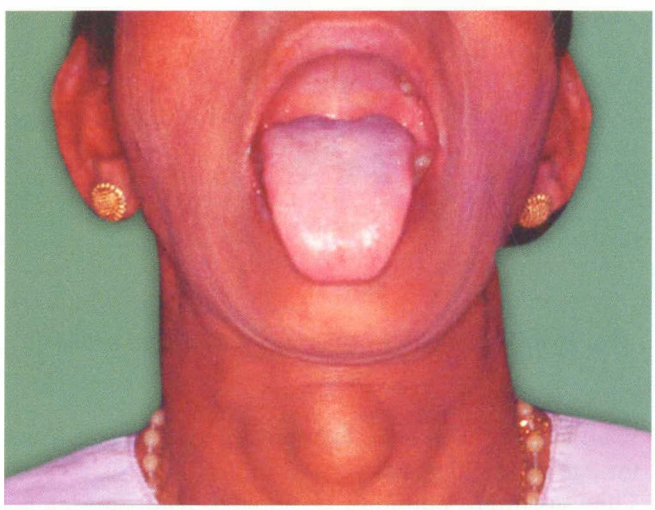


Fig. 16.47: Thyroglossal cyst: It moves upwards with protrusion of the tongue

KEY BOX 16.14

MOVEMENT ON PROTRUSION OF THE TONGUE

- The cyst is attached to the hyoid bone. Hence, it gives a classical TUG.
- Not always present, cyst below the thyroid cartilage—tug is absent.
- Better appreciated on holding the swelling.

Examination of thyroglossal cyst

- Cyst proper, mobility
- Base of the tongue to rule out lingual thyroid and lymph nodes.

PEARLS OF WISDOM

Do not forget to feel base of the tongue for lingual thyroid/ectopic thyroid. Whenever a patient has one congenital anomaly, examine thoroughly for 'more' associated anomalies.

THYROGLOSSAL FISTULA

Thyroglossal fistula is **never congenital**. It is always acquired due to the following reasons (Fig. 16.48 and Key Box 16.16):

1. Infected thyroglossal cyst rupturing into the skin.
2. Inadequately drained infected thyroglossal cyst.
3. Incompletely excised thyroglossal cyst.
   - **The track is lined by columnar epithelium.**

Clinical features

- Previous history of swelling in front of the neck, which is now **painful, red and ruptured resulting** in discharging pus. Once the pus is drained, the opening closes. However, after an interval of time, the 'pain and discharge' reappear.

Fig. 16.48: Thyroglossal fistula

KEY BOX 16.15

RECURRENT ABSCESS: RUPTURE FISTULA OR SINUS

- Thyroglossal fistula
- Osteomyelitis
- Stitch abscess
- Pilonidal sinus
- Median mental sinus
- Cold abscess
- Umbilical sinus

KEY BOX 16.16

THYROGLOSSAL FISTULA

- Always acquired
- Fistulous opening is in the midline
- Semilunar sign or hood sign
- It gets pulled up with protrusion of the tongue

KEY BOX 16.17

SISTRUNK'S OPERATION

- Fistula with **entire thyroglossal tract** is excised.
- **Central portion of the hyoid bone** and lingual muscle are removed.
- Removal is facilitated by **pressing the posterior 1/3rd of the tongue**.
- **Do not perforate** thyrohyoid membrane.
- **Incomplete removal results in recurrence.**

KEY BOX 16.18

THYROGLOSSAL DUCT ANOMALIES

- Lingual thyroid
- Levator glandulae thyroidae
- Ectopic thyroid tissue
- Thyroglossal cyst

- When there is **no infection, the fistula** discharges only mucus and the surrounding skin is normal. Infected fistulae are tender, discharging pus and the skin is red hot.
- Majority of the patients presenting are young in the age group of 10 to 20 years.
- **A fistulous opening in the centre of neck** which is covered by a hood of skin can occur due to increased growth of the neck, when compared to that of fistula. This is described as **semilunar sign or hood sign.**

Treatment

- Infection is controlled with antibiotics.
- Surgical excision should include the **fistula with removal of the entire tract up to the foramen caecum.** Otherwise, recurrence will occur.
- The **central portion of the hyoid bone is removed** due to close proximity of the fistula.
- An elliptical incision is preferred as it gives a neat scar.
- This operation is called **Sistrunk's** operation.

  See Key Box 16.17 for details about Sistrunk's operation.

ANOMALIES OF THYROGLOSSAL DUCT

- Thyroglossal duct extends from foramen caecum to thyroid cartilage.
- Various anomalies have been given in Key Box 16.18.
- However, thyroglossal cyst is common. Lingual thyroid and ectopic thyroid tissue are uncommon swellings.
- They have to be kept in mind as a differential diagnosis of the swellings in the midline of the neck.

SWELLING ARISING FROM ISTHMUS OF THE THYROID GLAND

Almost all the diseases of the thyroid gland result in enlargement of the isthmus. However, a solitary nodule and cysts can occur in relation to isthmus. The swelling moves with deglutition. However, it does not move on protrusion of the tongue.

PRETRACHEAL AND PRELARYNGEAL LYMPH NODES

These lymph nodes produce nodular swelling in the midline. One or two discrete nodes are palpable. They can enlarge due to the following conditions:

1. **Acute laryngitis:** The nodes are tender, soft.
2. **Papillary carcinoma of thyroid:** The nodes are firm without matting, with or without evidence of thyroid nodule.
3. **Carcinoma of the larynx:** The nodes are hard in consistency.
4. **In India, tuberculosis** should be considered as a possible diagnosis when other diseases are ruled out.

SWELLINGS IN THE SUPRASTERNAL SPACE OF BURNS

1. **Lipoma:** Soft and lobular, edge slips under the palpating finger.
2. **Sequestration dermoid cyst** is a midline, soft, cystic, fluctuant swelling.
3. **Gumma** produces a firm swelling with evidence of syphilis elsewhere in the body.
4. **Thymic swellings, an aneurysm** of innominate or subclavian artery are the other causes (Fig. 16.49).

Fig. 16.49: Aneurysm

CLINICAL NOTES

A lady of 65 years presented with swelling in the suprasternal space (Fig. 16.92). *Candidate gave the diagnosis of lymph node swelling—probably cold abscess. He failed. It had expansile pulsations. It was a case of aneurysm of innominate artery. M.S. exam case 2008, JNMC, Belgaum (Courtesy: Professor Ashok Godhi, HOD of Surgery)*

DIFFERENTIAL DIAGNOSIS OF LATERAL SWELLINGS IN THE NECK

Before we discuss the swellings in the lateral side of the neck, it is essential to know the various triangles in the neck. These are discussed below.

TRIANGLES OF THE NECK

Each side of the neck is a quadrilateral space subdivided by sternocleidomastoid into anterior triangle and posterior triangle. They are further subdivided as given below (Fig. 16.50).

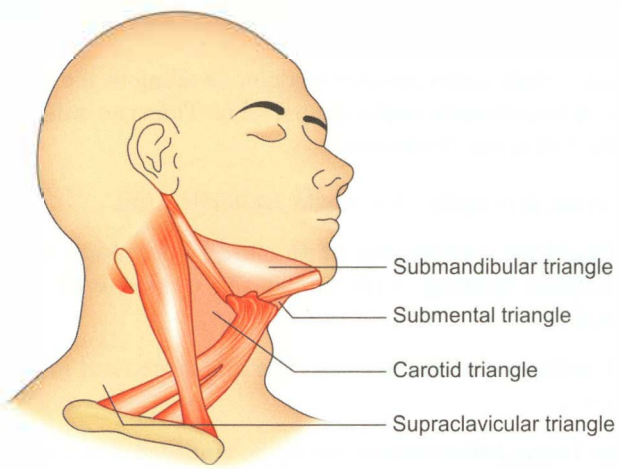


Fig. 16.50: Triangles of the neck

Anterior triangle

1. Submental triangle
2. Digastric (submandibular) triangle
3. Carotid triangle
4. Muscular triangle

Posterior triangle

1. Occipital triangle
2. Supraclavicular triangle

SWELLINGS IN SUBMANDIBULAR TRIANGLE

- The submandibular triangle is a part of anterior triangle.
- This is bounded **inferiorly** by anterior and posterior belly of digastric muscles with their tendon, **superiorly** by the attachment of deep fascia to the whole length of mandible
- This triangle is covered by deep fascia.
- The floor is formed by mylohyoid muscle which arises from mylohyoid line of the mandible, thus closing the space.
- **Swellings in the submandibular triangle are** (Fig. 16.51):
  1. Enlarged submandibular lymph nodes—common
  2. Submandibular salivary gland enlargement—common
  3. Plunging ranula—not uncommon
  4. Ludwig's angina—not uncommon
  5. Lateral sublingual dermoid cyst—rare
  6. Tumours of the mandible—rare

Fig. 16.51: This swelling in the submandibular triangle was bidigitally not palpable. However, it exhibited a doubtful sign of movement with deglutition. It turned out to be an ectopic thyroid swelling (*Courtesy:* Prof Sampath Kumar, Dept. of Surgery, KMC, Manipal)

ENLARGED SUBMANDIBULAR LYMPH NODES

They form a nodular swelling which is deep to deep fascia. They are palpable only in the neck (not intraorally). The nodes can get enlarged due to the following conditions:

1. **Acute lymphadenitis:** Very often, poor oral **hygiene** or a **caries tooth** produces painful, tender, soft enlargement of these lymph nodes. Extraction of the tooth or with improvement of oral hygiene, lymph nodes regress.
2. **Chronic tuberculous lymphadenitis** can affect these nodes along with upper deep cervical nodes. The nodes are firm and matted.
3. **Secondaries** in the submandibular lymph nodes arise from carcinoma of the cheek, tongue, palate. The nodes are **hard** with or without **fixity**.
4. **Non-Hodgkin's lymphoma** can involve submandibular lymph nodes along with horizontal group of nodes in the neck. The nodes are **firm** or **rubbery** in consistency.

SUBMANDIBULAR SALIVARY GLAND ENLARGEMENT (Key Box 16.19)

The various causes of submandibular salivary gland enlargement have been discussed under the salivary gland chapter. The common causes are chronic sialadenitis with or without a stone, tumours of the salivary gland or enlargement due to autoimmune diseases. They form irregular or nodular swelling. The diagnosis is confirmed by bidigital palpation of the gland. **Enlarged submandibular gland is bidigitally palpable because the deep lobe is deep to mylohyoid muscle.**

KEY BOX 16.19

SUBMANDIBULAR SALIVARY GLAND ENLARGEMENT

- Calculus
- Chronic sialoadenitis
- Cancer
- Chronic diseases: Autoimmune

DIFFERENTIAL DIAGNOSIS OF SWELLINGS IN THE CAROTID TRIANGLE

The carotid triangle has following boundaries (Fig. 16.52):

Laterally by sternomastoid muscle, superomedially by digastric muscle and stylohyoid muscle and inferomedially by omohyoid muscle.

Some important swellings in this triangle are as follows:

1. Branchial cyst
2. Lymph node swelling (cold abscess)
3. Aneurysm of carotid artery.
4. Enlargement of the thyroid gland
5. Carotid body tumour— rare
6. Laryngocoele—rare
7. Sternomastoid tumour—rare
8. Neurofibroma of the vagus

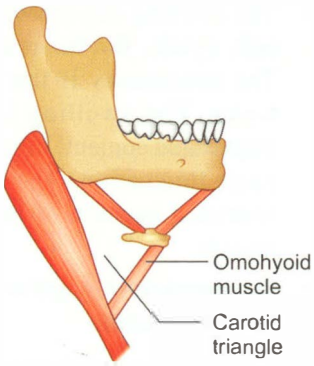


Fig. 16.52: Carotid triangle

BRANCHIAL CYST

Aetiology

- Branchial cyst arises from ***vestigeal remnants of 2nd branchial arch***.
- The cyst is lined by squamous epithelium and contains desquamated epithelial cells which slowly forms a tooth paste-like material.

Clinical features

- Even though congenital, majority of patients are young between the age group 15 and 25 years.
- The swelling is typically located in the **anterior triangle of the neck partly under cover of the upper 1/3rd of anterior border of sternomastoid**. This can be explained because of the development of *sternomastoid muscle from the myotome in the ridge of second branchial arch* (Fig. 16.53).

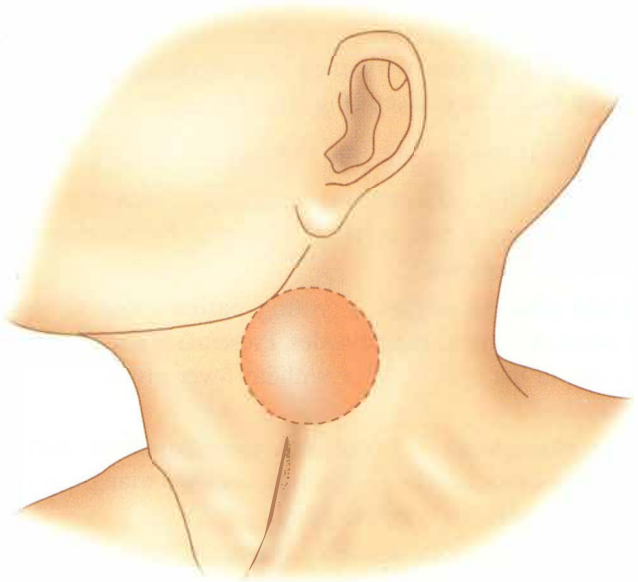


Fig. 16.53: Branchial cyst—typical location—partly covered by sternocleidomastoid muscle. Differential diagnosis for this swelling will be cold abscess

- The swelling has smooth surface and round borders. It is **soft, cystic, fluctuant and transillumination-negative**[1]. The consistency is that of a rubber bag half filled with water. The swelling is very often firm due to thick inspissated content. In such situations, it is very difficult to elicit fluctuation. The mobility of the swelling is also restricted because of its adherence to the sternomastoid muscle.
- **Sternomastoid contraction test:** The **swelling becomes less prominent.**
- If contents are aspirated, it contains **cholesterol crystals** (Key Box 16.20).
- No other lesion is found in the neck (lymph nodes).

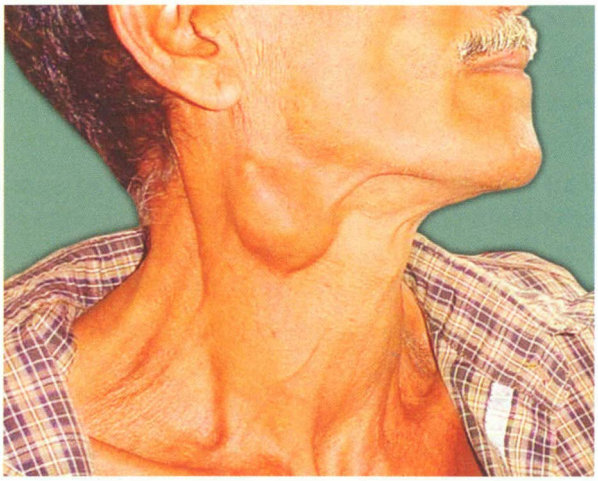


Fig. 16.54: Swelling mimicked branchial cyst. However, it was a subcutaneous swelling with lobularity—it turned out to be lipoma (*Courtesy:* Dr P Rajan, Calicut Medical College, Calicut, Kerala)

KEY BOX 16.20

SWELLINGS CONTAINING CHOLESTEROL CRYSTALS

- Branchial cyst
- Dental cyst
- Dentigerous cyst
- Hydrocoele

Treatment

- **Excision** of the cyst along with *its entire epithelial lining* with a curved incision centred over the swelling. One must ensure that epithelial lining should be removed completely or else recurrence will occur.
- Sometimes cyst may grow backwards in between 'fork' of common carotid artery as far as pharyngeal constrictors.

Complications

Since the wall is rich in lymphatic tissue, it can undergo secondary infection with pain and swelling. Hence, the swelling has to be excised (Fig. 16.54).

Differential diagnosis (Fig. 16.55)

There is no differential diagnosis in a classical case of branchial cyst. However, a few swellings have to be considered as differential diagnosis.

1. **Cold abscess** occurs in young patients due to tuberculosis of jugulodigastric nodes. Presence of multiple lymph nodes in the neck with or without fever gives clue to the diagnosis.
2. **Lymphangioma** is a brilliantly transilluminant, partially compressible swelling. However, anterior triangle is not a common site for lymphangioma.
3. **Lipoma** can also occur in the neck, though it is an uncommon site (Fig. 16.54).

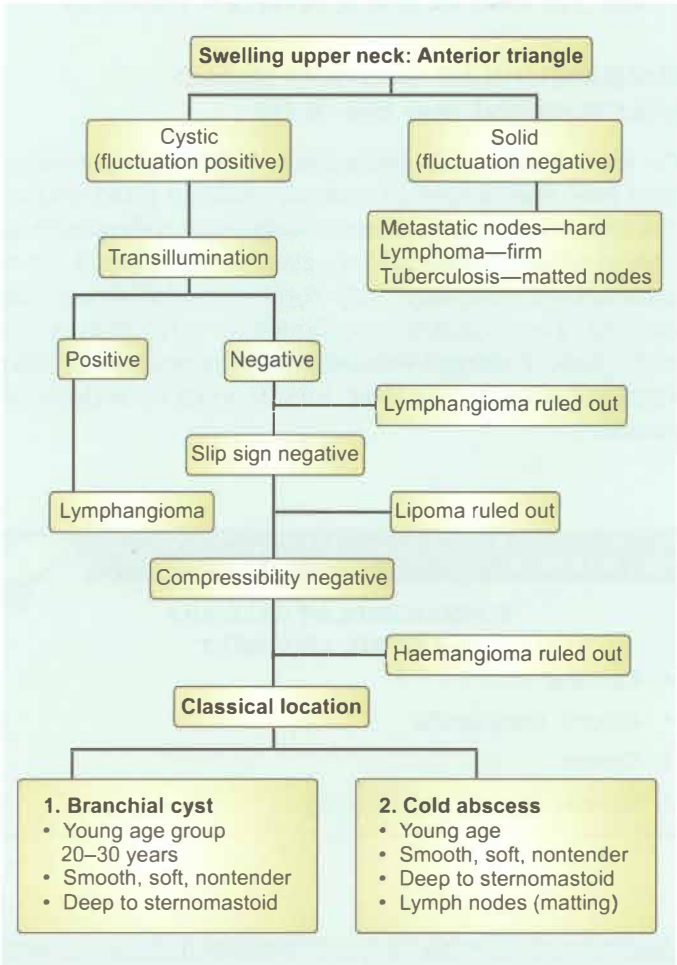


Fig. 16.55: Branchial cyst—one example of diagnosing a case using clinical methods. It is important that each clinical method **elicited carefully**, gives a clue to the diagnosis

[1]Branchial and thyroglossal cysts rarely give rise to transillumination if contents are clear and unless the cyst is small, it is difficult to elicit fluctuation. These cysts can be firm or hard swellings also.

BRANCHIAL FISTULA (Key Box 16.21)

- This is always congenital and occurs due to persistent **2nd branchial cleft.**[1]
- External opening is situated at the junction of middle 1/3rd and lower 1/3rd of sternomastoid (Fig. 16.56).
- The tract from the skin passes through the *fork of common carotid artery* deep to the accessory and hypoglossal nerve and opens in the **anterior aspect of posterior pillars of tonsils** (Fig. 16.56). The tract is lined by ciliated squamous epithelium and discharges a mucopurulent discharge. Sometimes, the upper end is blind resulting in a sinus.
- The patient may complain of a **dimple, discharging mucus** and the dimple becomes more obvious when the patient is asked to swallow.
- Usually seen in growing adults (30% of cases).
- Can be unilateral or bilateral, equally common in males and females.
- It is also called **lateral fistula of the neck.** (Thyroglossal fistula is called median fistula of the neck.)

Fig. 16.56: Sites of branchial fistula—commonly bilateral

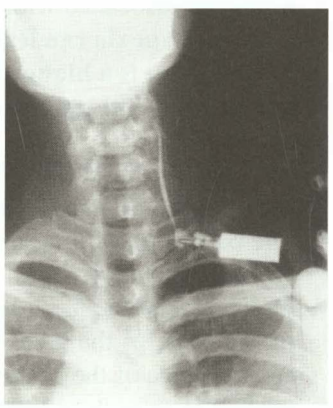


Fig. 16.57: Contrast study demonstrating fistulous tract

KEY BOX 16.21

BRANCHIAL FISTULA PASSES SUPERFICIAL TO FOLLOWING STRUCTURES

- Internal carotid artery
- Internal jugular vein
- Hypoglossal nerve
- Glossopharyngeal nerve
- Stylopharyngeous muscle

Derivatives of 3rd arch

Pierces superior constrictor muscle and opens on posterior pillar of the fauces behind tonsil.

[1]Persistent first branchial cleft results in external auditory meatus.

Treatment

- **Fistulogram** can be done by injecting methylene blue into the external opening and defining the tract (Fig. 16.57). This is followed by exploration of the tract. At surgery, it should be carefully dissected up to the internal opening and then excised. May have to be done by two different incisions: Upper incision at upper border of thyroid cartilage and lower incision encircling fistula and dissecting upwards.

Complication

Recurrent infection of the fistula.

COLD ABSCESS DUE TO TUBERCULOSIS

- In India, this is the commonest cystic swelling in the carotid triangle. The cold abscess occurs as a result of caseation necrosis of the lymph nodes. This forms a soft, cystic, fluctuant swelling with negative transillumination. Presence of other lymph nodes in the neck or sinuses in the neck gives the clue to the diagnosis.
- Loss of appetite, weakness and fever with chills may be other features.

ANEURYSM OF THE COMMON CAROTID ARTERY

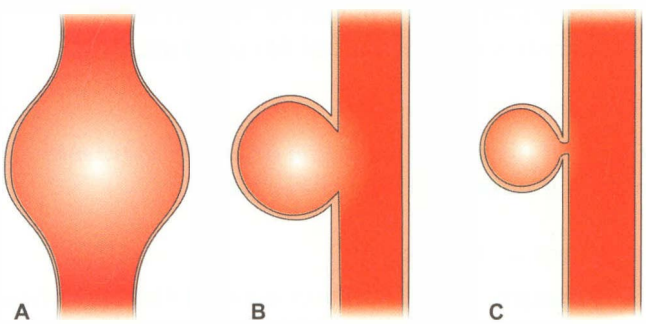
- ***Atherosclerosis*** is the most common cause of aneurysm. This weakens the vessel walls uniformly and produces fusiform dilatation of the blood vessel. ***Hypertension*** is another factor which adds to the aneurysm.
- ***Abdominal aorta*** is the commonest site for aneurysms followed by ***popliteal artery.***

Types

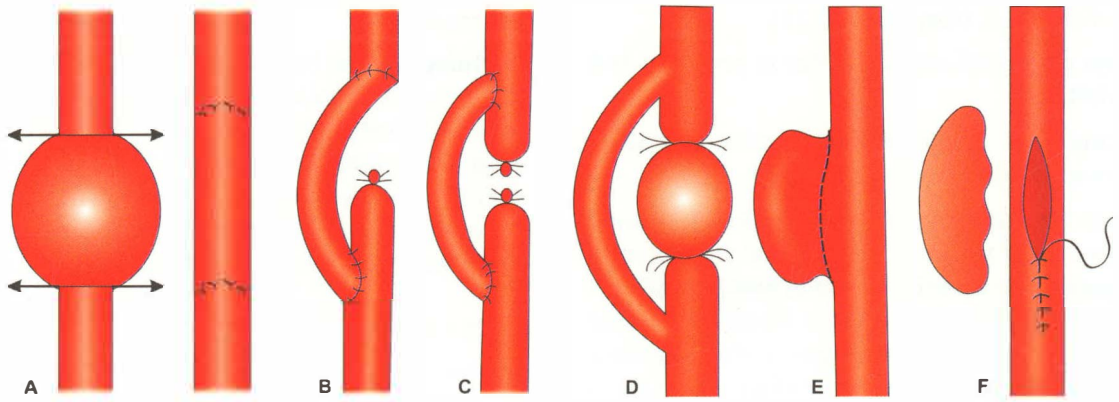
A. Fusiform: Atherosclerosis, hypertension (Fig. 16.58A).

B. Saccular: Due to injury (Fig. 16.58B).

C. False: In this condition there is a sac lined by cellular tissue which communicates with the artery through an opening in its wall (Fig. 16.58C).



Figs 16.58A to C: Fusiform, saccular, false aneurysm



Figs 16.59A to F: Different methods of operation for aneurysm. (A) Excision and end-to-end graft, (B) Excision and end-to-side graft, (C) Excision and side-to-side graft, (D) Excision and bypass grafting, (E and F) Matas aneurysmorrhaphy

Causes (Key Box 16.22)

KEY BOX 16.22

ANEURYSM: CAUSES

- **Congenital:** Berry aneurysm in the circle of Willis
- **Traumatic**
- **Degenerative:** Atherosclerosis
- **Rare causes:**
  Syphilis: Endarteritis obliterans
  Mycotic: Infective emboli
  Subacute bacterial endocarditis
  Marfan's syndrome
  Polyarteritis

Clinical features of aneurysm

1. Elderly patients are commonly affected.
2. Evidence of atherosclerosis in the form of thick walled vessel is present.
3. Tensely cystic (feels firm), fluctuant, transillumination negative swelling with ***expansile pulsation*** (when the fingers are kept over the aneurysm *they are not only elevated but they are separated*).
4. Compressibility is positive
5. On exerting pressure proximally the swelling diminishes in size—classically it happens in a case of popliteal aneurysms on compression of the femoral artery.
6. Bruit/thrill is characteristic of this condition.

PEARLS OF WISDOM

Classical signs of an aneurysm described above may be absent, if thrombus is present within it.

Treatment of aneurysm

- Angiography to confirm the diagnosis followed by repair of aneurysm with graft—PTFE graft (polytetrafluoroethylene graft, Fig. 16.59).

CAROTID BODY TUMOUR (CHEMODECTOMA)

Introduction

- This is a benign tumour arising from chemoreceptors in the carotid body (Key Box 16.23). They are situated in the tunica adventitia at the bifurcation of common carotid artery (Fig. 16.60).
- Hence, such a tumour is called chemodectoma.
- Function of the carotid body is regulation of pH.
- It may be associated with phaeochromocytoma.
- **Chronic hypoxia can lead to carotid body hyperplasia. Hence, there is a higher incidence of chemodectoma in people living at higher altitudes.**

Clinical features (Key Box 16.24)

- Middle-aged or elderly patients are affected (5th decade).
- The patient gives long history of painless, slow-growing swelling for many years.
- **Typical location:** In the upper part of the anterior triangle of the neck, at the level of the hyoid bone, beneath the anterior edge of the sternomastoid muscle (Fig. 16.61).
- Surface is smooth or lobulated, borders are round, and is an oval, vertically placed swelling. Consistency is firm to hard. Hence, called **classical potato tumour**.

Fig. 16.60: Location of carotid body

Fig.16.61: Carotid body tumour—classical site

KEY BOX 16.23

CHEMORECEPTORS: SITES

- The carotid body
- The aortic body
- Brainstem
- Pulmonary receptors
- Myocardial receptors

Hormonally not active cells

- **Horner's syndrome and unilateral vocal cord paralysis** can occur due to involvement of the nerves.
- Pressure on the tumour gives rise to syncopal attack due to decrease in the pulse rate **(carotid body syndrome)**
- Moves in the transverse direction.
- Carotid artery is stretched over the swelling and so, transmitted pulsations are felt (Fig. 16.62).
- Intra-oral examination shows prolapse of ipsilateral tonsil, unless it grows in parapharyngeal space.

Diagnosis

- Carotid angiography (Fig. 16.63A) should be done if there are neurological symptoms such as syncopal attack. It may demonstrate separation of the carotid bifurcation.
- **Lyre sign:** Splaying of carotid artery can be seen (Fig. 16.63B)
- Incision biopsy is dangerous
- Colour Doppler should be the first investigation.

Treatment

- Excision of the tumour with reconstruction
- No role for radiotherapy.

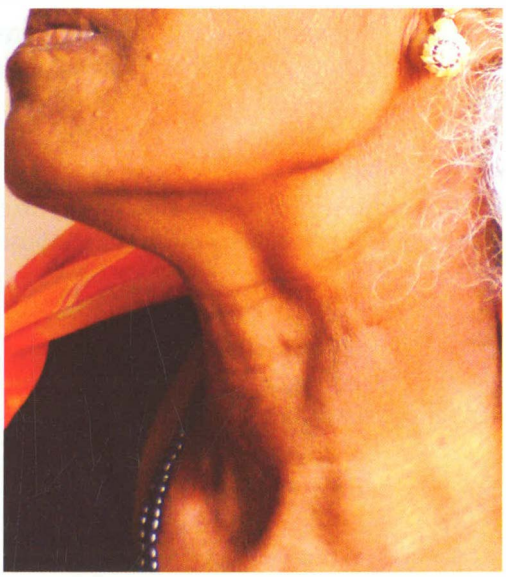
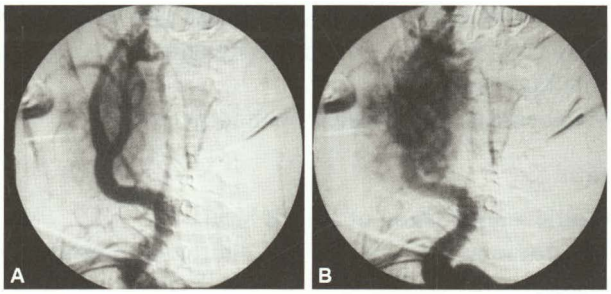


Fig. 16.62: Carotid body tumour in a 45-year-old lady—duration 5 years (*Courtesy:* Dr MR Srivatsa, Prof and Head, Prof Bagali Babasaheb, Prof Bharathi, Dr Srikar Pai, Department of Surgery, MS Ramaiah Medical College and Hospital, Bangalore)



Figs 16.63A and B: Carotid angiography—observe separation of internal and external carotid artery by the tumour (A) and vascular blush (B)

KEY BOX 16.24

CAROTID BODY TUMOUR

- Rare tumour
- Rarely malignant
- Rarely bilateral
- Rarely grows fast
- Rarely patient presence early
- Rarely metastasises
- Experience of a general surgeon with this tumour is very, very rare.

PEARLS OF WISDOM

It is important to preserve cerebral circulation during surgery. Vascular surgeon's help is necessary.

Complications

- Very rarely, it can turn into a malignant carotid body tumour with lymph nodal metastasis.

Precautions

- Do not biopsy from within mouth—carotid body tumour can displace tonsil medially.
- Should not do FNAC
- Should not do open biopsy
- When you feel some pulsations over a lymph node in the carotid triangle—remember carotid body tumour.

STERNOMASTOID TUMOUR

- This is not a tumour, it is a **misnomer**.
- Injury to the sternomastoid during birth causes rupture of few fibres and ***haematoma.*** Later, healing occurs with fibrosis, resulting in a swelling in the middle of sternomastoid muscle.
- The other possible theory is that this is a congenital anomaly—**short sternomastoid muscle**.

Clinical features

- This is seen in infants or children. Firm to hard, 1–2 cm swelling in the middle of the sternomastoid muscle.

- Tender and mobile sideways. Medial and lateral borders are distinct but superior and inferior borders are continuous with the muscle.
- Many cases are associated with torticollis.

Treatment

- Gentle manipulation of child's head
- Physiotherapy to stretch the shortened sternomastoid muscle.
- Division of lower attachment of sternomastoid from clavicle and sternum with or without removal of lump is the surgical treatment.

LARYNGOCOELE (Key Box 16.25)

- It occurs due to herniation of the laryngeal mucosa (Fig. 16.64, external laryngocoele)
- When it enlarges within the larynx, it may displace vocal cord, produce hoarseness and is called **internal laryngocoele.**

Causes

- Glass blowers, musicians, wind instruments and trumpet players are commonly affected.
- Chronic cough may be one of the predisposing factors.

Clinical features

- Smooth, oval, boggy swelling which moves upwards on swallowing, in relation to thyrohyoid membrane (subhyoid position).
- Swelling becomes prominent when the patient is asked to cough or blow ***(Valsalva manoeuvre).***

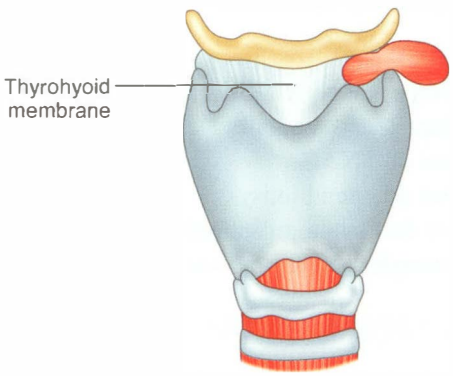


Fig. 16.64: Laryngocoele

KEY BOX 16.25

LARYNGOCOELE

- Very rare
- Increased laryngeal pressure
- Expansile impulse on cough
- Treatment: Ligation of its neck and division of the whole sac.

- Expansile cough impulse is present.
- Tympanitic note on percussion (resonant)

Treatment

- Excision of the sac—in external laryngocoele
- Marsupialisation—in internal laryngocoele.

Differential diagnosis

Other cystic swellings such as branchial cyst and lymphangioma should be ruled out.

Complications

Secondary infection results in laryngopyocoele. The opening in the thyrohyoid membrane may be blocked by mucopus in such cases.

PHARYNGEAL POUCH

- Herniation or protrusion of mucosa of the pharyngeal wall through Killian's dehiscence.
- **Killian's dehiscence** is a potential area of weakness in between the two parts of the inferior constrictor muscle (Fig. 16.65): **(A)** Upper oblique fibres (thyropharyngeus) and **(B)** Lower horizontal fibres (cricopharyngeus).

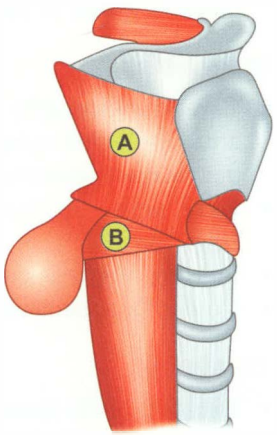


Fig. 16.65: Pharyngeal pouch

Aetiopathogenesis

Due to increase in the intrapharyngeal pressure, mucous membrane bulges in between parts of inferior constrictor muscles due to neuromuscular imbalance. Hence, it is a pulsion diverticulum.

Course of the diverticulum

Pulsion diverticulum deviates to one side mostly to the left because of the rigid vertebral column in the midline posteriorly.

Diagnosis

- Initially foreign body sensation is present in the throat. Later, gurgling sound regurgitation of food on turning to one side, sense of suffocation, cough or dysphagia is present.
- Aspiration may cause dyspnoea later.

PEARLS OF WISDOM

Pharyngeal pouch is a swelling behind sternocleidomastoid below the level of thyroid cartilage—soft swelling which can be emptied.

Treatment

- Barium swallow followed by excision of the pouch
- Cricopharyngeal myotomy may also be done.

SCHWANNOMA OF THE VAGUS NERVE (Fig. 16.66)

- This condition produces swelling in the carotid triangle in the region of thyroid swelling.
- It is a vertically placed oval swelling
- It is firm to hard in consistency
- On pressure over the swelling, dry cough and in some cases bradycardia may occur.

Fig. 16.66: Schwannoma of the vagus nerve (*Courtesy:* Prof P Rajan, Calicut Medical College, Kerala. This swelling was not moving with deglutition—Candidate offered thyroid nodule as diagnosis—MS exam case 2007)

DIFFERENTIAL DIAGNOSIS OF SWELLING IN THE POSTERIOR TRIANGLE

The posterior triangle (Key Box 16.26) is an interesting area as far as swellings are concerned. It is the commonest area of metastasis in lymph nodes from occult primary. Lymphangiomas, haemangiomas, cold abscess, lymphomas commonly occur here. Interesting cases of cervical rib, Pancoast's tumour, aneurysms also occur here.

KEY BOX 16.26

BOUNDARIES OF POSTERIOR TRIANGLE

- Anteriorly : Sternomastoid (posterior border)
- Laterally : Trapezius (anterior border)
- Above : Mastoid process
- Below : Clavicle

Most of the swellings have been discussed under appropriate chapters. Haemangioma, metastasis in the cervical lymph nodes and Pancoast's tumour have been discussed below.

Classification (Table 16.5)

Common swellings in the posterior triangle are given below.

HAEMANGIOMA

Definition

This is a swelling due to congenital malformation of blood vessels. It is an example of ***Hamartoma***.

Classification

A. Depending on the origin

- Capillary
- Cavernous
- Arterial

B. Depending on behaviour of the lesion

	Terms in common use
I. Involuting haemangioma	Strawberry naevus
• Superficial	Capillary haemangioma
• Deep	Cavernous
• Combined (superficial + deep)	Strawberry naevus
	Capillary haemangioma
	Cavernous haemangioma
II. Noninvoluting	
• Portwine stain	Portwine stain
	Capillary haemangioma
	Naevus flammeus
• Cavernous haemangioma	
• Arteriovenous fistula	

Table 16.5 Swellings in the posterior triangle

I. Solid swellings	II. Cystic swellings	III. Pulsatile swellings
1. Metastasis in the lymph nodes	Lymphangioma	Subclavian artery aneurysm
2. Tuberculosis	Haemangioma	Vertebral artery aneurysm
3. Lymphoma	Cold abscess	
4. Lipoma		
5. Cervical rib		
6. Pancoast's tumour		

CAPILLARY HAEMANGIOMA (Key Box 16.27)

They consist of dilated capillaries and proliferation of endothelial cells. Hence, it commonly occurs in the skin (Figs 16.67 and 16.68). They can be of the following types:

1. **Salmon patch** is a bluish patch over the forehead, in the midline, present at birth and disappears by 1 year of age. Hence, no treatment is required.
2. **Port-wine stain** is an extensive intradermal haemangioma. This is bluish purple in colour, commonly affects the face or other parts of the skin, is present at birth and usually progresses and does not regress (Key Box 16.28).
   - It is a noninvoluting capillary haemangioma (dilatation due to defective maturation of cutaneous innervations during embryogenesis).
   - Area supplied by sensory branches of the fifth cranial nerve is involved.
   - Start with light red colour and progress to deep colour.
   - Pulsed dye laser using light with specific wavelength of 585 or 595 nanometres is **one of the best treatments** available. This process is called as **'photothermolysis'.**
   - It may be associated with **Sturge-Weber syndrome** (page 259)

PEARLS OF WISDOM

Flat, patchy lesion on the face in the area of fifth cranial nerve that does not fade is port-wine stain.

KEY BOX 16.27

CAPILLARY HAEMANGIOMA

- Skin and soft tissue involvement
- Salmon patch: Midline–forehead
- Port-wine stain: Head and neck
- Strawberry angioma: Compressible
- Wait and watch policy is the best.

KEY BOX 16.28

PORT-WINE STAIN

- **P**ort-wine colour even though to start with it is red in colour
- **O**ccurs usually in the face, can also occur on the shoulder and trunk
- **R**egression does not occur
- **T**reatment: Pulsed dye laser, photocoagulation, dermabrasion
- **W**orrying because it becomes more keratotic and nodular as age advances
- **I**njection of sclerosants may be needed
- **N**oninvoluting haemangioma is its another name
- **E**xtensive intradermal capillary dilatation

Remember as **PORT-WINE**

3. **Strawberry angiomas** produce swelling which protrud from the skin surface. The child is normal at birth. After month, a ***bright red swelling*** appears over the head an neck region, which exhibits ***sign of compressibility***. Th lesion consists of immature vascular tissue. Even thoug the lesion grows initially, by 5–7 years of age, ***swellin regresses and colour fades.*** Hence, no specific treatmen is necessary. The treatment is indicated only when th swelling persists. 70% resolve by 7 years of age.

VENOUS (CAVERNOUS) HAEMANGIOMA

This occurs in place where venous space is abundant, e.g. lip cheek, tongue, and posterior triangle of the neck (Fig. 16.6 and Key Box 16.29).

Clinical features

- History of a swelling in the neck of long duration. History of bleeding is present when it occurs in the oral cavity.
- The swelling is warm and ***bluish in colour*** but not pulsatile.
- Soft, fluctuant, transillumination is negative.
- **Compressibility** is present. This sign is also called 'sign of emptying' or 'sign of refilling'. When the swelling is compressed between the fingers, blood diffuses under the vascular spaces and when pressure is released, it slowly fills up. Compressibility is a diagnostic sign of haemangioma.

Differential diagnosis

1. **Lymphangioma is brilliantly transilluminant.** If a lymphangioma is infected or has been treated with preliminary injections, it may not show transillumination.
2. **Lipoma** is not compressible.
3. **Cold abscess**
4. **Branchial cyst** when it is in anterior triangle.

Fig. 16.67: Capillary haemangioma (*Courtesy:* Prof Pramod Kumar, HOD of Plastic Surgery, KMC, Manipal)

KEY BOX 16.29

CAVERNOUS HAEMANGIOMA

- Compressible swelling
- Bluish warm, nontender swelling
- Associated with arteriovenous communication
- Associated with lipoma: Naevolipoma

CLINICAL NOTES

Gluteal region is one of the sites of haemangioma, AV fistula, aneurysm and neurilemmoma. A patient presented with this swelling which was firm with a few cystic areas. It was excised in toto. Remember to check whether it is a low pressure (venous) or a high pressure arterial lesion before excising a haemangioma. Massive bleeding can occur if it is a high pressure lesion.

Treatment of cavernous haemangioma

Principles

1. **Injection** is the first line of treatment of cavernous haemangioma. It makes the swelling **fibrotic, less vascular and small**. Thus, excision can be done at a later date (Key Boxes 16.30 and 16.31).
2. **Excision** of haemangioma in the oral cavity is more difficult than in the neck.
3. It is better to have a control of external carotid artery in the neck, while excising haemangioma in the oral cavity. If necessary, external carotid artery should be ligated in order to control the bleeding.
4. Adequate blood to be arranged.
5. **Previous embolisation** into the feeding artery decreases the size of the haemangioma (therapeutic embolisation).

KEY BOX 16.30

INJECTION LINE OF TREATMENT

- Boiling water, hypertonic saline or sodium tetradecyl sulphate (STD solution) can be used.
- In multiple spaces, in multiple sittings.
- Obliteration occurs due to aseptic thrombosis and fibrosis.
- Lesion becomes flat.

KEY BOX 16.31

SWELLINGS: TREATED WITH SCLEROSANTS

- Haemangioma
- Haemorrhoids
- Prolapse rectum
- Oesophageal varices
- Varicose veins

6. ***Large haemangiomas in the oral cavity*** should be excised only after ***preliminary sclerotherapy*** and taking all the precautions mentioned above.

Syndromes associated with haemangioma
(Table 16.6, Figs 16.76 and 16.77)

Complications of haemangioma (Key Box 16.32)

KEY BOX 16.32

COMPLICATIONS OF HAEMANGIOMA

- Ulceration and bleeding: Commonly occurs with capillary haemangioma
- Infection: Septicaemia usually precipitated by a small ulcer
- High output cardiac failure

CONGENITAL ARTERIOVENOUS (AV) FISTULA
(Arterial Haemangioma)

- An abnormal communication between artery and vein, results in AV fistula (Key Box 16.33).
- AV fistula can be congenital or acquired (Figs 16.73, 16.79 and 16.80).
- Such AV fistula has got structural and functional effects (Figs 16.78A and B)

Structural effect

- Since high pressure blood from an artery flows into the vein, the veins get dilated, tortuous and elongated. This arterialisation of the vein results in secondary varicose veins.

Table 16.6 Syndromes associated with haemangioma and its findings

Syndromes associated with haemangioma	Findings
1. Klippel-Trenaunay-Weber syndrome	1. Naevus flammeus, osteohypertrophy of extremities, AV fistula and varicose veins
2. Osler-Rendu-Weber syndrome	2. Haemangioma of lip associated with haemangioma of GIT
3. Sturge-Weber syndrome	3. Haemangioma of brain, mental retardation, Jacksonian epilepsy, glaucoma

Fig. 16.68: Capillary haemangioma of the nose

Fig. 16.69: Cavernous haemangioma of the cheek

Fig. 16.70: Local gigantism due t haemangioma involving gluteal region

Fig. 16.71: Sign of compressibility was positive in this case. It was initially diagnosed as post-auricular dermoid cyst

Note: Importance of complete clinical examination

Fig. 16.72: Haemangioma tongue causing macroglossia. She also had haemangioma of the cheek and skin

Fig. 16.73: AV malformation since birth and growing since 3 years. The patient had bleeding due to trivial trauma. Local rise of temperature and machinery murmur was present

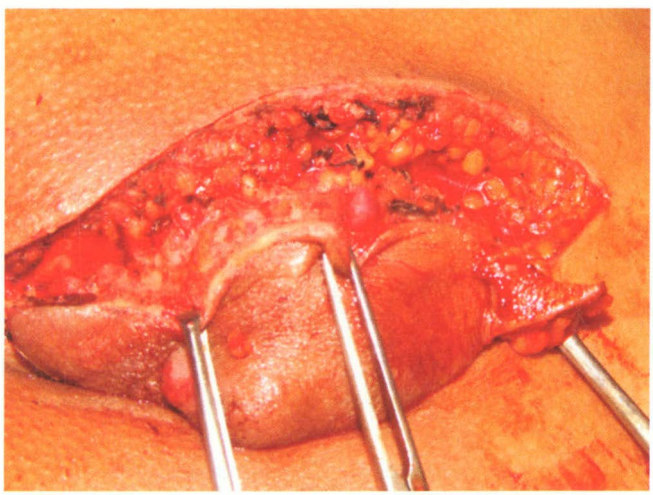


Fig. 16.74: AV malformation being excised

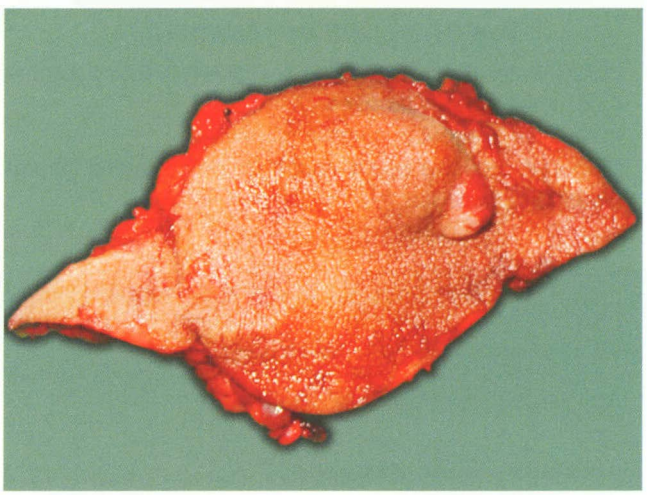
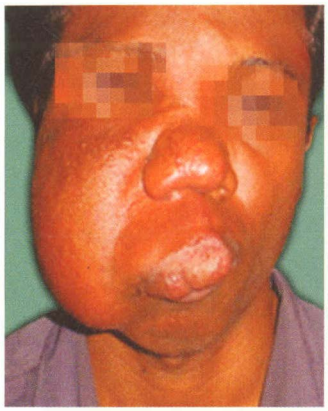
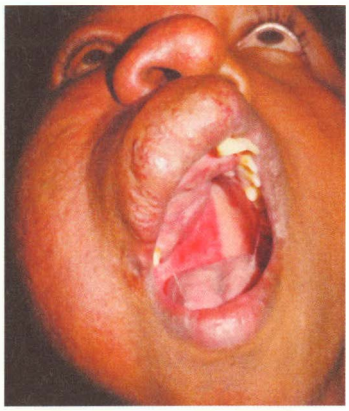


Fig. 16.75: Sclerosants were injected initially to reduce vascularity. This was followed by wide excision

Figs 16.76 and 16.77: Large haemangioma involving cheek, lip and palate. He also had haemangioma of the liver

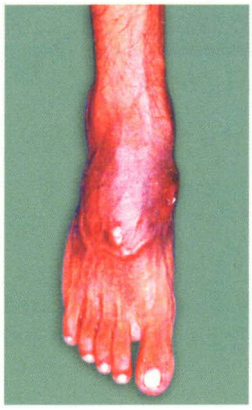


Fig. 16.78A: Traumatic AV fistula. *See* the arterialisation of the vein

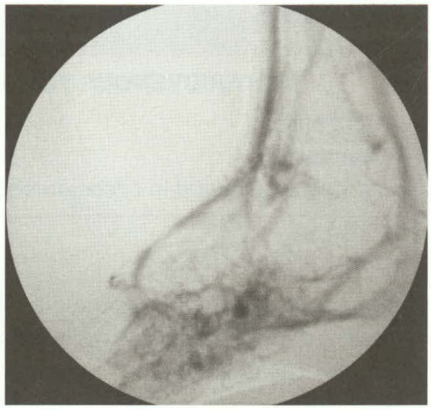


Fig. 16.78B: DSA picture

Fig. 16.79: Most common type of AV fistula you see today is in the nephrology ward—created to facilitate haemodialysis

Fig. 16.80: X-ray of the hand—traumatic AV fistula

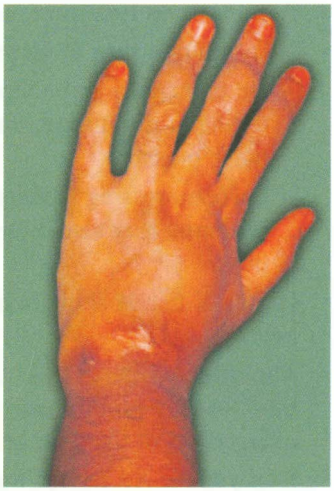


Fig. 16.81: Traumatic AV fistula affecting wrist and hand. Kindly observe prominent veins and swollen fingers

Fig. 16.82: Congenital AV fistula of 25 years duration

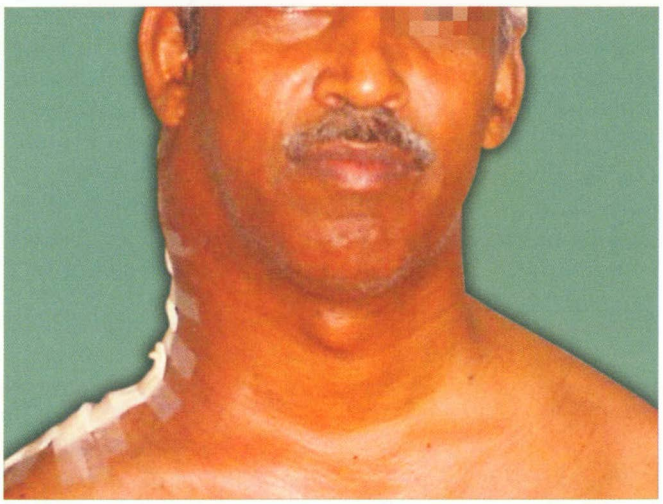


Fig. 16.83: After therapeautic embolisation it has regressed by over 70%. Patient is waiting for another course of embolisation. (*Courtesy:* Dr Umesh Bhat, Surgeon, Kundapur and Dr Subhaschandra, Interventional cardiologist, Manipal Hospital, Bangalore)

KEY BOX 16.33

ARTERIOVENOUS FISTULA: TYPES

- Congenital
- Traumatic
- Iatrogenic: Created in cases of renal failure

Physiological effect

- Increased pulse rate, increased cardiac output, increased pulse pressure result due to increased venous pressure and arteriovenous shunt.

Functional effect

- Soft, cystic, fluctuant, transillumination—negative, **pulsatile swelling.**
- A continuous bruit/murmur is characteristic (Fig. 16.73)
- **Nicoladoni's sign or Branham's sign**
  - On compressing the feeding artery, the venous return to the heart diminishes, resulting in fall in pulse rate and pulse pressure.
  - On compressing feeding artery, pulsation or **continuous murmur** may also disappear and swelling will diminish in size.
- If the AV fistula is big, a **high output cardiac failure** can occur.
- The affected part is swollen (because of high pressure) than—**local gigantism** (Figs 16.70 and 16.72). Thus, overgrowth of the limb or toe can occur.
- Distal to the AV fistula, there are **ischaemic ulcers**, due to comparative reduction in the blood supply.

CLINICAL NOTES

This lady visited many general practitioners for a swelling near the coccyx region (adjacent to natal cleft). The diagnosis which was made in this case was subcutaneous neurofibroma, pilonidal sinus abscess, boil and haemangioma. Careful examinations revealed pulsations. History dates back to 20 years duration. It was a case of congenital AV fistula. Wide excision was done. She has been asymptomatic for 2 years now. [Courtesy: Dr B Hartimat, Asst. Prof of Surgery, KMC, Manipal (Figs 16.74 and 16.75)*].*

CLINICAL NOTES

A 30-year-old patient who had an injury to the dorsum of hand, had pain and swelling in dorsum of hand for 60 days duration. Many had missed the diagnosis. However, swelling had local rise of temperature, pulsations and reducibility (Fig. 16.81).

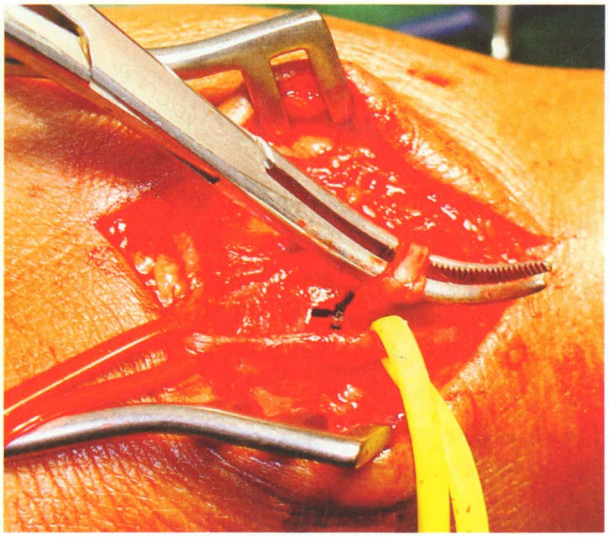


Fig. 16.84: Traumatic AVM at surgery

Investigations

- Angiography with **DSA** pictures (digital subtraction angiography) are essential before treating these patients (Fig. 16.78B).

Treatment

- Therapeutic embolisation is the treatment of choice for arteriovenous fistula, in congenital cases (Figs 16.82 and 16.83).
- Acquired lesion need to be observed or treated by quadruple ligation (Fig. 16.84).

CIRSOID ANEURYSM

- Not an aneurysm
- It is an AV fistula occurring in older people affecting the temporal region.
- The arteries and the veins are dilated and tortuous and are compared to pulsating bag of worms.

COLD ABSCESS IN THE POSTERIOR TRIANGLE (Fig. 16.85)

Causes

1. **Posterior cervical lymph nodes** primarily involved—route of infection from **adenoids** or other lymph nodes in the anterior triangle.
2. Lower posterior lymph nodes or Scalene node—route of infection from **lungs**.

Fig. 16.85: Cold abscess in the posterior triangle

3. From tuberculous cervical spine: Caries spine

- Clinically it presents as pain in the back, cold abscess and neurological presentation.
- **Rust's sign:** Child with caries spine will support the head by holding the chin.
- Cold abscess from caries spine can rupture anteriorly or posteriorly.

A. Anterior rupture: It ruptures deep to prevertebral layer of deep cervical fascia. From here, it can take the following routes:

- **Upper cervical region:** Presents as deep seated abscess in the posterior wall of the pharynx in the **midline**.
- **Lower cervical region:** Pus will press on oesophagus and trachea forwards.
- **Laterally** pus passes deep to prevertebral **fascia behind carotid sheath** in the **posterior triangle** (Fig. 16.86).

B. Posterior rupture

- Pus may enter spinal canal and then can travel along anterior primary division of the cervical spinal nerves.

Diagnosis

- **Cervical spine X-ray** to rule out spinal tuberculosis.
- **Chest X-ray** to rule out pulmonary tuberculosis.
- **Nondependent aspiration of the cold abscess** followed by AFB staining.

Treatment

- Antituberculous treatment
- Nondependent aspiration if cold abscess is present.
- Please refer orthopaedic books for specific treatment of TB spine.

Differential diagnosis

- Haemangioma—compressible
- Lymphangioma—transilluminant
- Schwannoma (Fig. 16.86)

Fig. 16.86: Schwannoma in the posterior triangle. Soft tissue sarcoma is another diagnosis. Undergraduate clinics at JNMC, Belgaum, Karnataka (*Courtesy:* Dr Aditya Patil, Prof. Ashok Godhi, HOD of Surgery)

SECONDARIES IN THE LYMPH NODES OR LYMPH NODE METASTASIS IN HEAD AND NECK

Introduction

Very often, the patients present to the surgeon with lymph node swelling in the neck with or without any complaints. If there is an obvious lesion in the oral cavity, the diagnosis is easy. On the other hand, difficulty arises in locating the primary malignancy, which is hidden or occult. It is important to know the anatomical location of the lymph nodes in the neck and drainage area, so that drainage areas can be investigated.

Classification

- They can be divided into horizontal (Table 16.7) and vertical groups of lymph nodes. Interestingly, very often circular group of nodes are enlarged in non-Hodgkin's lymphoma on both sides.

Table 16.7 Horizontal group of lymph nodes

Location	Drainage area	Common diseases
1. Submental	Central part of the lower lip, floor of the mouth, tip of the tongue	Infections, metastasis
2. Submandibular	Cheek, tongue, whole of the upper lip, outer part of the lower lip, gums, angle of mouth, side of nose, inner angle of the eye	Infections, carcinoma
3. Parotid	Nasopharynx, back of nose, eyelids, front of scalp, enamel, auditory meatus, tympanic cavity	Eyelid tumours, parotid tumours, tuberculosis
4. Preauricular	Outer surface of pinna, side of scalp	Scalp infection, tuberculosis
5. Postauricular	Temporal region of scalp, back of pinna	Scalp infection
6. Occipital	Back of the scalp	Scalp infection, secondary syphilis

VERTICAL GROUP OF LYMPH NODES (Fig. 16.87)

They can be classified into central and lateral group of lymph nodes. Most of the central group of lymph nodes are in relation to laryngeal cartilages or the trachea. The lateral lymph nodes are the most popular lymph nodes called deep cervical nodes. Majority of them are in close relationship with internal jugular vein. Jugulo-omohyoid and jugulodigastric lymph nodes belong to this category.

The details of the vertical group of lymph nodes are given in Table 16.8.

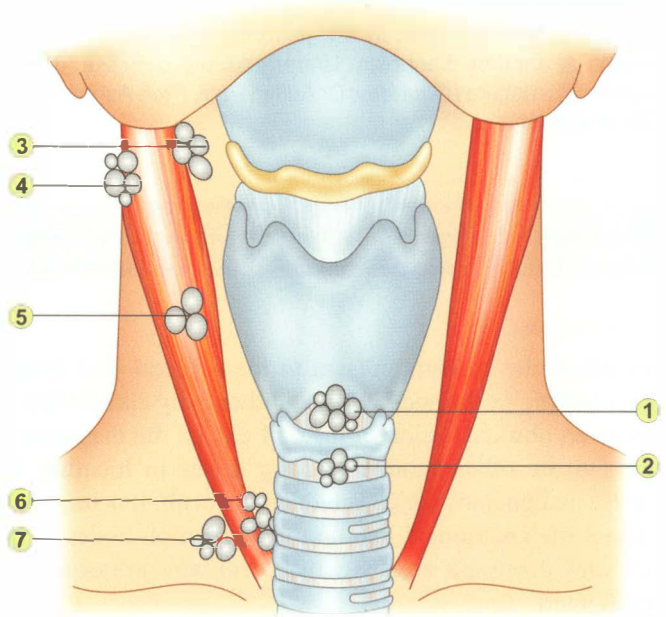


Fig. 16.87: Vertical group of lymph nodes (*see* Table 16.8)

Memorial Sloan-Kettering cancer centre: Latera lymph node classification

Level I : Lymph nodes in the **submental triangle anc submandibular triangle.**

Level II : **Upper jugular nodes** from posterior belly o digastric superiorly to hyoid bone inferiorly.

Level III : **Middle jugular nodes** from hyoid superiorly tc cricothyroid membrane inferiorly.

Level IV : **Lower jugular nodes** from cricothyroic membrane superiorly to clavicle inferiorly.

Level V : **Posterior cervical region** from anterior borde of trapezius posteriorly to posterior border of sternocleidomastoid anteriorly and clavicle inferiorly.

Level VI : **Anterior compartment nodes** from hyoid bone superiorly to suprasternal notch inferiorly and laterally by medial border of carotid sheath.

Level VII : **Upper mediastinal nodes** inferior to suprasternal notch (no longer used).

- It is advisable to know these various levels of lymph nodes and their drainage areas.
- When block dissection is being done, you will find the lymph nodes being mentioned in the form of levels.

Factors which accelerate local and regional spread of malignant tumour (Key Box 16.34).

KEY BOX 16.34

FACTORS WHICH ACCELERATE LOCAL AND REGIONAL SPREAD OF MALIGNANT TUMOUR

- Angiogenesis
- Lymphangiogenesis
- Lack of basement membrane on lymphatic vessels
- Extracapsular breakout

Table 16.8 Vertical group of lymph nodes

Location	Drainage area	Common diseases
1. Prelaryngeal (on the cricothyroid ligament)	Larynx	Laryngeal carcinoma
2. Pretracheal, paratracheal (in front of trachea)	Trachea, thyroid	Papillary carcinoma thyroid, tuberculosis
3. Upper anterior deep (jugulodigastric)	Tonsil, posterior 1/3 tongue, oropharynx, pyriform recess	Tonsillitis, carcinoma posterior 1/3 tongue, oropharyngeal carcinoma, tuberculosis
4. Upper posterior deep	Adenoids, posterior pharynx, retropharyngeal region	Tuberculosis, nasopharyngeal carcinoma
5. Middle group of lymph nodes	Thyroid, supraglottis	Papillary carcinoma thyroid
6. Lower anterior (jugulo-omohyoid)	Tongue, thyroid, subglottis	Carcinoma tongue, carcinoma thyroid
7. Lower posterior (supraclavicular)	Thyroid, postcricoid, oesophagus, lungs breast	Bronchogenic carcinoma, intra-abdominal malignancy, lymphoma

Note: True vocal cords are devoid of lymphatics.

- Example: In papillary carcinoma thyroid, level III, level IV, level V and level VI lymph nodes are removed (*see* next page).

Clinical presentation of metastatic deposits in the lymph nodes

1. Majority of patients are elderly males (> 50 years), present with painless swelling in the neck of a few months duration.
2. The symptoms with which a patient presents to the hospital gives the clue to the site of origin of the primary. Few examples are given below:
   - **Difficulty in swallowing:** Carcinoma posterior 1/3 tongue, oropharyngeal carcinoma or carcinoma oesophagus
   - **Difficulty in breathing:** Laryngeal cancer
   - **Hoarseness of voice:** Larynx or thyroid
   - **Obvious growth in oral cavity:** Carcinoma cheek, alveolus, tongue, etc.
   - **Haemoptysis, difficulty in breathing:** Bronchogenic carcinoma
   - **Epistaxis, ear pain or deafness:** Nasopharyngeal carcinoma (Key Box 16.35)

Clinical signs

- Lymph nodal metastasis appears as a **hard, nodular** or irregular mass in the anatomical location of the lymph nodes.
- Early cases may have some mobility. However, in majority of cases **nodes get fixed** and they attain a huge size. Very often, what appears as one lymph node, is a complex mass of multiple lymph nodes.
- On sternomastoid contraction test or chin test, these nodal swellings become less prominent.
- Skin ulceration is a late feature. **A prominent skin fold** is due to infiltration into the platysma—**platysma sign**.
- The primary malignancy may be evident in the anterior third of tongue, cheek, alveolus, etc.
- Posterior one-third of the tongue should be palpated with gloved finger (Key Box 16.36).

KEY BOX 16.35

NASOPHARYNGEAL CARCINOMA[1]: TROTTER'S TRIAD

- Conductive deafness
- Homolateral immobility of soft palate
- Pain in the side of the head due to involvement of 5th cranial nerve

KEY BOX 16.36

OCCULT PRIMARY SITES

- Posterior 1/3rd of the tongue, oropharynx
- Nasopharynx, sinuses
- Upper oesophagus, bronchus, thyroid

- Secondaries in the lymph nodes can cause pressure effects or may cause paralysis of nerves. Upper anterior deep cervical lymph nodes can cause **hypoglossal nerve paralysis** (Key Box 16.37) where in the tongue points towards the side of lesion. **When there is no evidence of the primary lesion clinically, the situation is described as occult primary** with secondaries in the neck.
- Pain in the distribution of trigeminal nerve (face) suggests nasopharyngeal malignancy infiltrating skull base (foramen lacerum).

How do you suspect metastasis in the neck?

Any elderly patient presenting to the hospital with firm to hard lymph node in the neck of short duration with or without fixity. He has no signs and symptoms of inflammation such as fever or pain to begin with.

Having suspected a metastatic deposit, remember the following facts:

- **80%** of them are **metastatic** deposits.
- Majority of malignant neoplasms are **epithelial** in origin.
- Nodes in the **upper half** (Level I and II) can be due to **primary in the oral cavity, tongue, oropharynx, larynx.**
- Nodes in the **lower half** (Level III and IV) **can be due to primary in the thyroid, tongue.**
- **Nodes in the supraclavicular region** (Level V): Carcinoma in the GIT, genitourinary tract, lungs and nasopharynx.
- **Nodes in the pretracheal, suprasternal region** (Level VI): Papillary carcinoma thyroid.

KEY BOX 16.37

CLINICAL EXAMINATION IN A CASE OF LYMPH NODES IN THE NECK

- Lymph nodes: All groups
- Drainage areas

Pressure effects on:

- Hypoglossal nerve
- Accessory nerve
- Cervical sympathetic chain

[1]More details about nasopharngeal carcinoma are given in page 291 in the chapter on oral cavity.

METASTASIS IN CERVICAL LYMPH NODES—VARIOUS LEVELS (Figs 16.88 to 16.96)

Fig. 16.88: Level I and II nodes. Carcinoma floor of the mouth

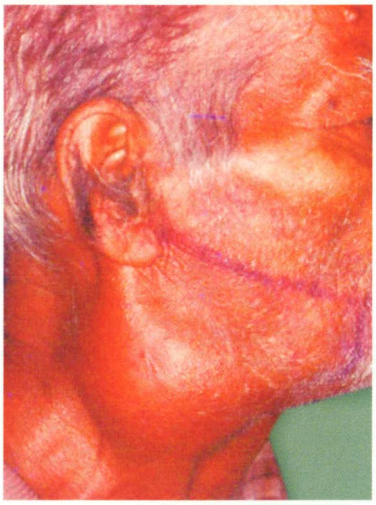


Fig. 16.89: Level II and III nodes: Carcinoma alveolus

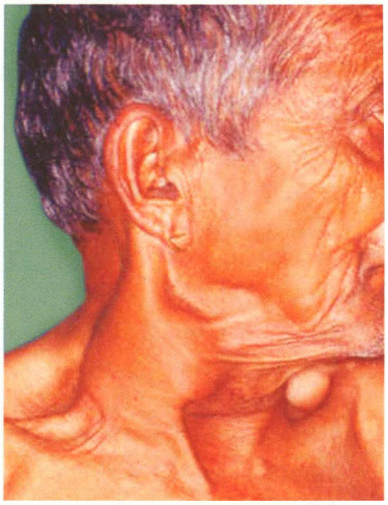


Fig. 16.90: Level III, IV and V nodes Carcinoma posterior 1/3rd of tongue

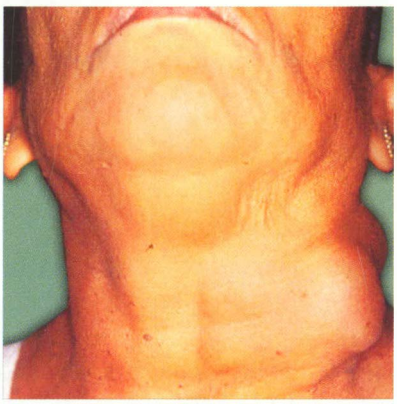


Fig. 16.91: Level V nodes: Post-cricoid carcinoma

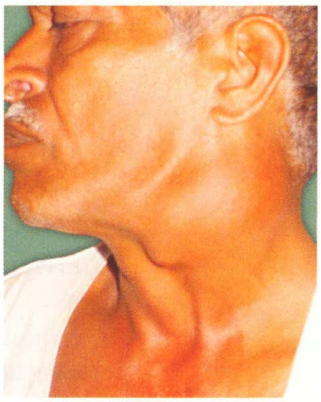


Fig. 16.92: Predominantly Level II, III and V nodes: From nasopharyngeal carcinoma

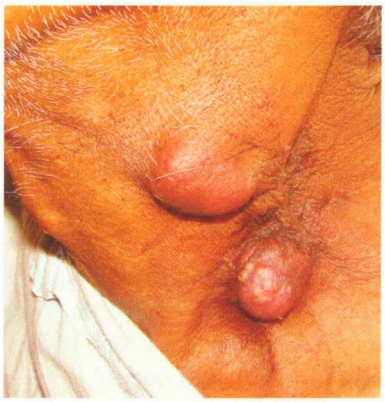


Fig. 16.93: Predominantly Level II, III and V nodes: From nasopharyngeal carcinoma

Fig. 16.94: Level I, II, III and V nodes: Primary could not be identified—a case of occult primary

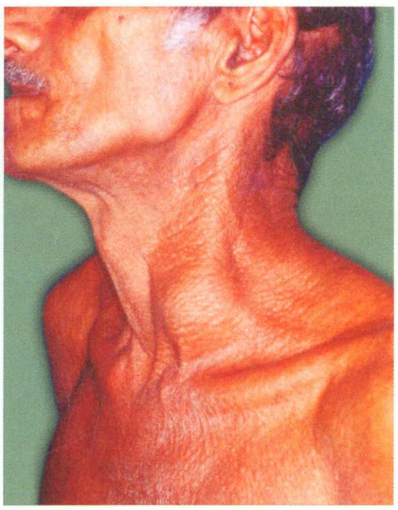


Fig. 16.95: Scalene node: In between the two heads of sternomastoid—a case of bronchogenic carcinoma

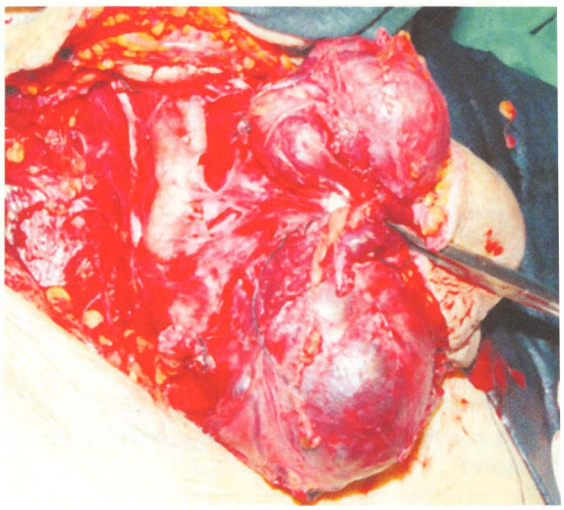


Fig. 16.96: Hugely enlarged lymph nodes which are bluish in colour, pushing the carotid arteries anteriorly—papillary carcinoma thyroid

NECK DISSECTION (Fig. 16.103)

Introduction

- **Crile** in 1906, first described a standardised dissection of cervical nodes by applying **anatomical and oncological principles** including removal of surrounding fibrofatty tissue from various compartments of the neck.
- Radical neck dissection causes significant functional and cosmetic morbidity.
- Today adjuvant chemoradiotherapy is an important modality of treatment.
- Thus with better understanding of tumour biology, natural history of disease and availability of adjuvant radiotherapy, radical neck dissection is not **frequently done but its modifications are done.**
- Rule of 80 in neck masses (Key Box 16.38)

Indication

- Carcinoma tongue, carcinoma floor of mouth
- Malignant melanoma
- Metastatic lymph nodes from pharynx and upper oesophagus.

Contraindications

- Fixed nodes, evidence of distant metastasis
- Untreatable primary cancer

Types (*see* next page)

I. Classical radical neck dissection (Crile's operation)

Removal of level I to V nodes + IJV, sternomastoid + spinal accessory and submandibular salivary gland + Tail of parotid.

Incision

1. MacFee: Two incisions are given (Fig 16.97)

- Upper incision extends from mastoid process to hyoid bone up to the point of chin across intermediate tendon of digastric muscle.
- Lower incision is given 2 cm above clavicle—from anterior border of trapezius to midline.
- Gives a very good exposure and vascularity of flaps are good. No corners and hence, no necrosis.

2. Crile's incision

- Upper incision is similar to MacFee. The other incision is oblique along the length of sternocleidomastoid inclining more in the posterior triangle (Fig. 16.98).
- When sternocleidomastoid has to be removed as a part of neck dissection or to clear the **lymph nodes which** are badly **stuck to jugular vein or to accommodate PMMC flap, Crile's is a better incision** (Fig. 16.99).
- Here all the cervical lymph nodes from Level 1 to Level VI are removed **along with nonlymphatic structures such as** sternocleidomastoid muscle, internal jugular vein, accessory nerve (XI), submandibular salivary gland and cervical sympathetic plexus. A few examples wherein radical neck dissection is done are **carcinoma tongue, oropharyngeal** carcinoma, etc.

II. Modified radical neck dissection (MRND—Fig. 16.100)

Type I

- *Preserve one structure:* Spinal accessory nerve
- Classically done for squamous cell carcinoma of upper aerodigestive tract with clinically positive neck dissection.

Type II MRND

Preserve two structures: Spinal accessory and IJV.

Type III MRND or BOCCA's functional neck dissection (Fig. 16.101)

- *Preserve three structures:* Spinal accessory, sternocleidomastoid and internal jugular vein.
- Done for metastatic well differentiated carcinoma thyroid.
  In this all the lymph nodes from Level I to Level V are removed but **nonlymphatic structures are preserved.**
- Dissection is from lower border of mandible to clavicle and from anterior border of trapezius to midline.

III. Selective neck dissection

- Here any of the lymphatic compartments is preserved (which should have been removed as part of classic RND). A few examples are given below

a. Supraomohyoid dissection (Fig. 16.102): Removal of nodes in Level I, II and III is done for carcinoma oropharynx, carcinoma cheek. Usually done for carcinoma floor, lateral tongue, etc.

b. Lateral neck dissection: Level II, III, IV are removed as in carcinoma larynx and cervical oesophagus.

c. Posterolateral neck dissection: Level II to V are removed as in cutaneous malignancy of posterior scalp and neck. Can also be done for thyroid malignancies.

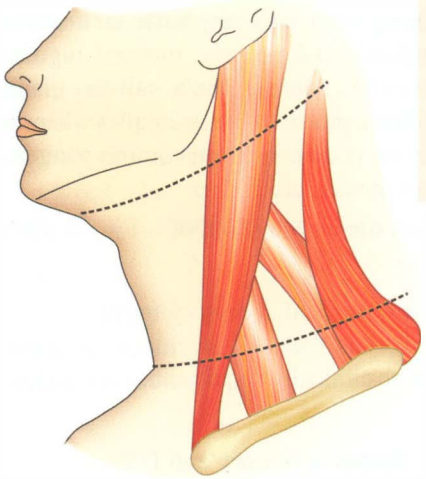
IV. Commando's operation

RND, hemimandibulectomy with radical glossectomy. It is a very radical and aggressive surgery done for carcinoma tongue.

KEY BOX 16.38

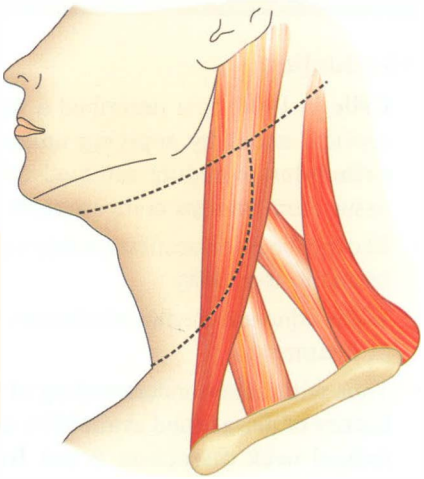
RULE OF 80 IN NECK MASSES

- 80% of neck masses are neoplastic
- 80% of neoplastic masses occur in males
- 80% of neck masses are malignant
- 80% of malignant neck masses are metastatic
- 80% of metastatic neck masses are from primary sites above clavicle.



- Double transverse incision
- 3 point intersection is avoided
- Subsequent blow outs of carotid artery can be avoided

- Furcate incision—otherwise called Y incision
- 3 point intersection occurs which may give rise to necrosis



Figs 16.97 and 16.98: Incisions for block dissection: MacFee and Crile

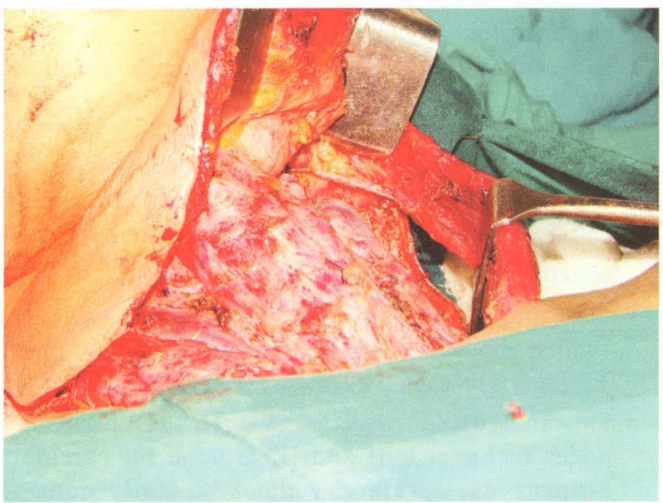


Fig. 16.99: Sternomastoid retracted

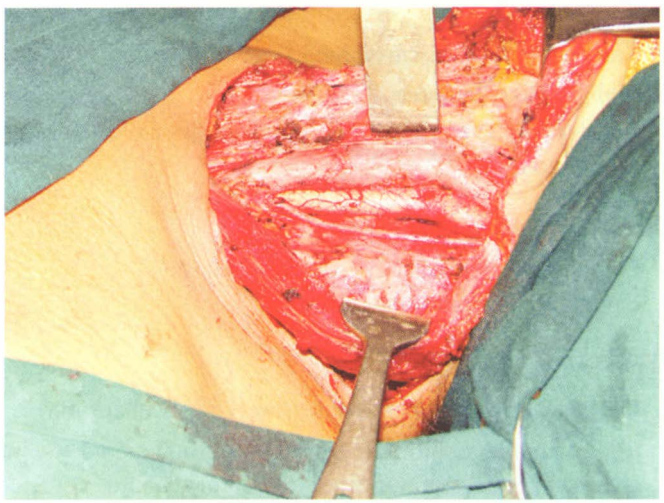


Fig. 16.100: Radical neck dissection

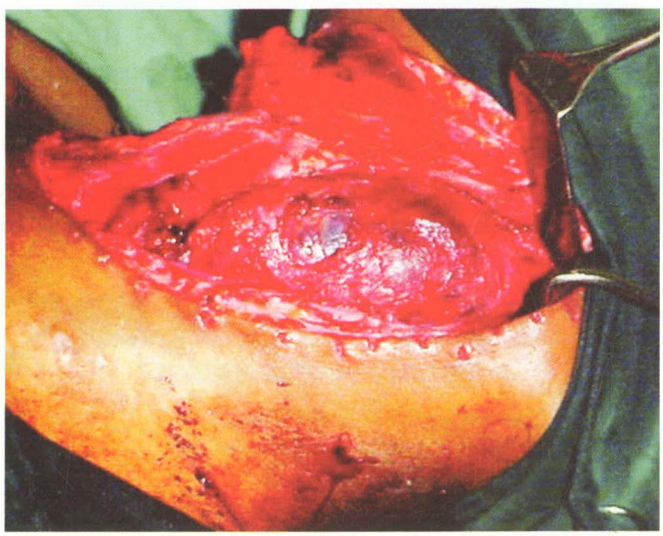


Fig. 16.101: Functional neck dissection—blue nodes of papillary carcinoma thyroid are unmistakable

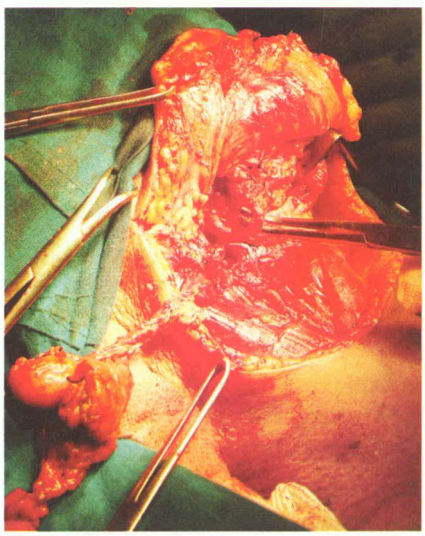


Fig. 16.102: Supraomohyoid neck dissection

Investigations

1. **Complete blood picture**
2. **Chest X-ray** can provide the following information:
   - Secondaries in the lungs with cannonball appearance as in cases of malignant melanoma of head and neck.
   - Bronchogenic carcinoma can be suspected by an irregular dense shadow in the peripheral lung fields.
   - Large mediastinal node mass may be seen, with or without tracheal shift.
3. **Biopsy** from clinically obvious lesion (tongue, cheek, etc.).
4. **Triple endoscopy includes**
   - Direct and indirect laryngoscopy
   - Oesophagoscopy
   - Bronchoscopy and biopsy of the suspicious area.
5. **X-ray base of the skull** may show destruction of the bone by the tumour.
6. **CT scan** of the sinuses, or nasopharyngeal area, or skull base to detect a primary growth, its extension, etc.
7. **FNAC** of the lymph nodes can give a diagnosis is more than 90% of the cases, **avoid incision biopsy as it will result in tumour recurrence and wound necrosis**.
8. If primary tumour cannot be detected on endoscopy, a **blind biopsy** is taken from **posterior wall of the fossa of Rosenmüller** and of the **pyriform fossa** on the same side.
9. When aspiration cytology is negative, an excision biopsy is advised as a last resort.

Treatment

1. **When the primary tumour is obvious** with enlarged lymph node metastasis, radiotherapy is the preferred line of treatment. However, *see* Chapter 17 for management of oral malignancy.
2. **Treatment of occult primary with metastatic lymph nodes**
   - If the lymph node or nodes are mobile, radical block dissection is done. If histopathologic report detects poorly differentiated carcinoma, extracapsular spread, or nodes are large (> 3 cm), radiotherapy is given.
   - If there are large, bulky, bilateral secondaries or fixed secondaries, radiotherapy is given to the neck masses. If the nodes do not completely regress or the tumour becomes bigger in size after 6–8 weeks of radiotherapy, radical neck dissection would be considered.

Follow-up

- About 30–40% of treated patients with occult primary with metastatic nodes die with no evidence of the primary later.
- In about 30% of patients, the primary will manifest within 1–2 years time.
- About 10% of patients are **cured** but primary is not detected.

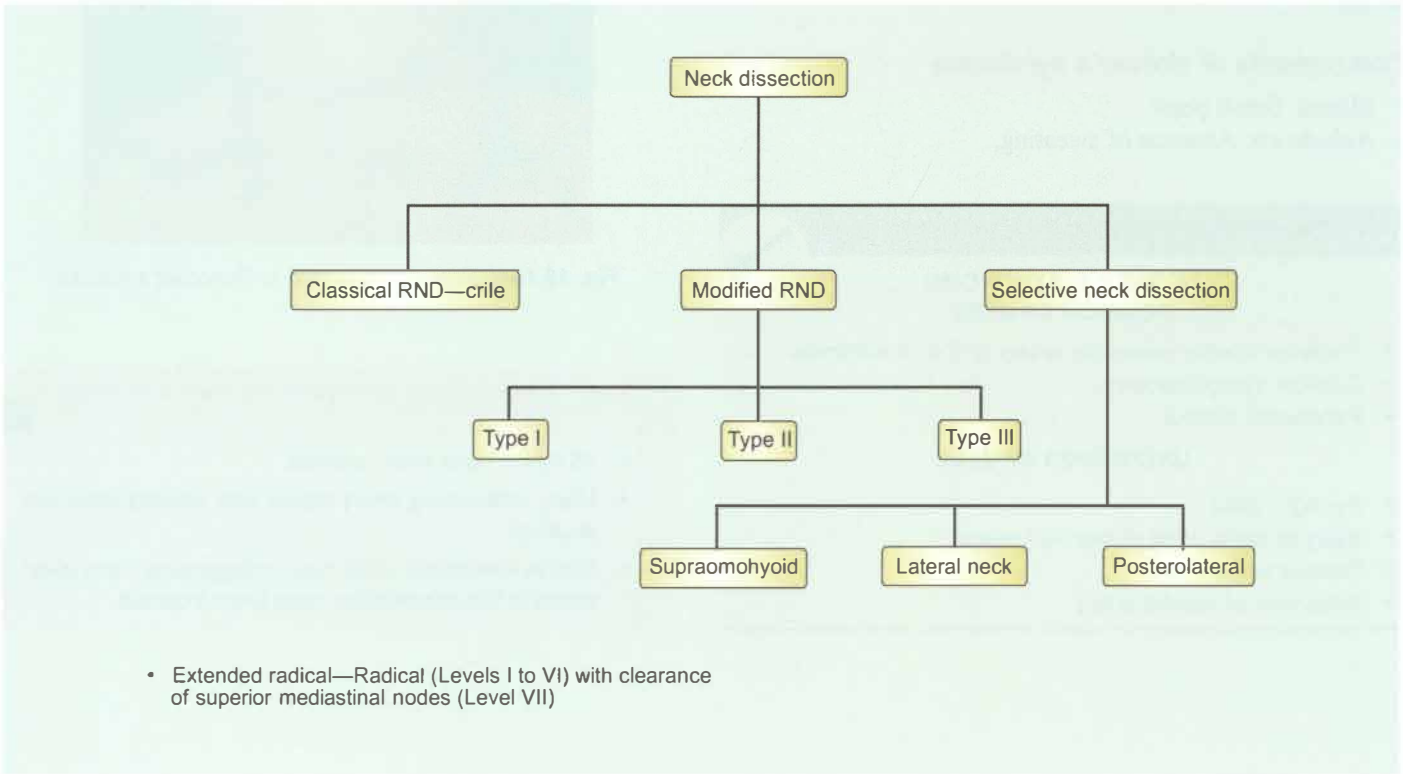


Fig. 16.103: Types of radical neck dissection

PANCOAST'S TUMOUR

- **Pancoast's tumour** or superior sulcus tumour is a bronchogenic carcinoma arising from the apex of lung.
- Typically, the patient is an elderly male around 70 years, **chronic smoker** who presents with cough, weight loss, dyspnoea and chest pain.
- As the tumour grows, it compresses the **lower roots of brachial plexus C8 and T1** and results in tingling, pain and paraesthesia in the distribution of ulnar nerve.
- The tumour is felt in the lower part of the posterior triangle. It is **hard in consistency, fixed, irregular** and sometimes tender. The lower border of the mass cannot be appreciated.

The Pancoast's syndrome refers to the following components:

1. Pancoast's tumour
2. Erosion of the first rib
3. Paralysis of C8 and T1 nerve roots
4. **Horner's syndrome** due to paralysis of cervical sympathetic chain. The preganglionic sympathetic fibres of the head and neck are given from the 1st and sometimes the 2nd thoracic segments of the spinal cord. These nerve fibres synapse with the cells in the three cervical sympathetic ganglia. They give rise to postganglionic fibres to the head and neck region.

Thus, anywhere along this pathway, disruption, damage or infiltration of the nerve roots results in Horner's syndrome. The causes of Horner's syndrome are depicted in Key Box 16.39.

Components of Horner's syndrome

- Miosis: Small pupil
- Anhidrosis: Absence of sweating.
- Pseudoptosis: Drooping of upper eyelid (Fig. 16.104).
- Enophthalmos: Regression of the eyeball
- Nasal vasodilatation: Nasal congestion

KEY BOX 16.39

HORNER'S SYNDROME
COMMON CAUSES

- Posterior inferior cerebellar artery (PICA) thrombosis
- Cervical sympathectomy
- Pancoast's tumour

UNCOMMON CAUSES

- Syringomyelia
- Injury to lower roots of brachial plexus
- Tumour in the neck
- Aneurysm of carotid artery

Investigations

1. **Chest X-ray:** May demonstrate a dense mass or collapse of the lobe, etc.
2. **CT scan** may demonstrate infiltration of the tumour into ribs or vertebra.
3. **Sputum for malignant cells**
4. **Flexible bronchoscopy:** Tissue biopsy or sputum sample can be collected.
5. **FNAC of the tumour** gives the diagnosis in majority of cases.

Treatment

Palliative radiotherapy. The response rate is poor.

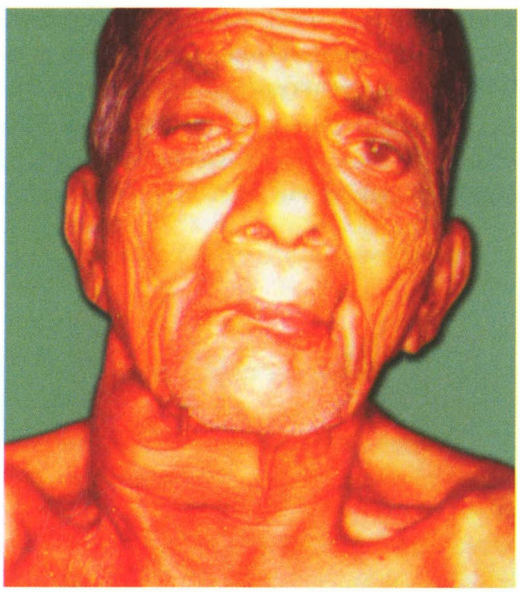


Fig. 16.104: Pseudoptosis due to Pancoast's tumour

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated.
- Many interesting short cases and photographs are inserted.
- Simple flow charts which help in diagnosing many short cases in the examination have been included.

MULTIPLE CHOICE QUESTIONS

1. Following are true for ranula *except:*
A. It is a swelling in the floor of the mouth
B. It is a retention cyst
C. It is transilluminant
D. Plunging ranula produces one more swelling in the submental region

2. Which one of the following does not give rise to cross fluctuation:
A. Iliopsoas abscess
B. Compound palmar ganglion
C. Sebaceous cyst
D. Hydrocoele *en bisac*

3. Following are true for thyroglossal fistula *except:*
A. Always congenital
B. It is lined by columnar epithelium
C. Semilunar sign is seen in adults
D. Surgery done for this is called Sistrunk's operation

4. Following are derived /arises from 2nd branchial arch *except:*
A. Sternomastoid muscle
B. Branchial cyst
C. Facial muscles
D. Anterior belly of the digastric

5. Which one of the following swelling does not contain cholesterol crystals?
A. Branchial cyst
B. Sebaceous cyst
C. Dental cyst
D. Hydrocoele

6. Most important stimulus for carotid body tumour is:
A. Hypoxia
B. Hyperbaric oxygen
C. Hypothermia
D. Hypercarbia

7. Tuberculous cervical spine can give rise to cold abscess in following locations *except:*
A. Posterior wall of the pharynx in the midline
B. Behind the carotid sheath
C. Front of the carotid sheath
D. Along the anterior primary division of the cervical spinal nerves

8. Pancoast' tumour has following features *except:*
A. It is a superior sulcus tumour
B. It can given rise to Horner's syndrome
C. It can erode first rib
D. It is ususually resectable

9. Components of Horner's syndrome are following *except:*
A. Miosis
B. Anhydrosis
C. Pseudoptosis
D. Exophthalmos

10. Following organs drain to posterior triangle lymph nodes
A. Adenoids
B. Thyroid
C. Retropharynx
D. Tonsil

11. Content of sebaceous cyst includes
A. Desquamated epithelial debris
B. Keratin
C. Sebum
D. Pus

ANSWERS

1 D 2 C 3 A 4 D 5 B 6 A 7 C 8 D 9 D 10 D
11 B

17

Oral Cavity, Odontomes, Lip and Palate

- Oral cancer
- Premalignant conditions
- General principles
- Carcinoma of buccal mucosa
- Carcinoma of tongue
- Ulcers of tongue
- Carcinoma of lip
- Carcinoma maxillary antrum
- Nasopharynx—cancer
- Benign lesions in the oral cavity
- Odontomes
- Epulis
- Median mental sinus
- Vincent's angina
- Cleft lip and cleft palate
- Ectopic salivary gland tumour
- Mucous cysts
- What is new ?/Recent advances

Introduction

Oral cavity is bounded by the lips anteriorly, the cheek on each side, tonsils posteriorly, superiorly by the palate and inferiorly by the floor of the mouth (Key Box 17.1). It is lined by squamous epithelium. Oral cavity is a common site of malignancy because it is insulted by various agents such as alcohol, smoking, tobacco chewing. **Oral cancer is the commonest malignant neoplasm** in the **head and neck**. Hence, it is discussed first in this chapter.

Abuse of tobacco and alcohol are the most common preventable risk factors for development of head and neck cancers (Key Box 17.2). Those who smoke 2 packs/day and drink 4 units of alcohol/day have an odds ratio of 35 for development of carcinoma. Tobacco quid is very dangerous and highly carcinogenic.

An interaction between **redox-active metals in saliva** and **low reactive free radicals in cigarette smoke occur**. Net result is that saliva loses its **antioxidant** capacity and instead becomes a potent pro-oxidant milieu.

Field cancerisation is a concept based on prolonged exposure of oral and pharyngeal mucosa to carcinogens. 15 to 20% of the survivors of one cancer of head and neck develop another primary head and neck cancer.

KEY BOX 17.1

ORAL CAVITY SUB SITES

- Lip
- Buccal mucosa
- Lower alveolus
- Retromolar trigone
- Oral tongue
- Floor of mouth
- Upper alveolus
- Hard palate

KEY BOX 17.2

RISK FACTORS ASSOCIATED WITH CANCER OF HEAD AND NECK

- **T**obacco quid[1]—'Pan Masala'
- **O**ropharyngeal Ca—**Plummer-Vinson syndrome**
- **B**arr-Epstein virus
- **A**lcohol, areca nut
- **C**igarette smoking and reverse smoking[2]
- **C**hronic irritation—Dentures
- **O**ral hygiene poor and poor nutrition

Remember as **TOBACCO**

[1]**Betel nut** chewed in combination with lime and cured tobacco is called **'quid'**. It is commonly placed in gingivolabial sulcus. **It is highly carcinogenic.** This type of carcinoma is very common in India called Indian oral cancer.

[2]**Reverse smoking:** Smoking a cheroot with burning end inside mouth. The risk of hard palate carcinoma is 47 times more in these patients.

ORAL CANCER

Incidence of oral cancer

- Tongue : 50%
- Cheek : 20–25%
- Floor : 10–15%
- Gums : 10%

Definitions

- **Hyperkeratosis** refers to increase in keratin layers. It occurs due to **constant irritation**. Once the cause is removed, it is **reversible**. It is a microscopic diagnosis.

  For example, Smokers' hyperkeratosis of the palate and lips. Once the aetiological agent is withdrawn, the lesion returns back to normal.
- **Leukoplakia** appears clinically as a white patch in the mouth and cannot be scraped off. It is irreversible and not attributable to any known disease. It is important to biopsy leukoplakic portion to rule out malignancy.

PEARLS OF WISDOM

Sump area or 'Coffin corner' at the posterior tongue/floor of mouth is a common site for cancer—may be missed on cursory inspection.

PREMALIGNANT CONDITIONS FOR ORAL CANCER

1. Leukoplakia

The causes for leukoplakia are as follows:

- **Smoking** results in hyperkeratosis. Nicotine in the form of cigarettes, chewed tobacco[1], powdered snuff produces premalignant changes in the oral cavity.
- **Spices**
- **Spirits** have synergistic action with smoking
- **Sharp** tooth, **sepsis**, poor oral hygiene
- **Sunlight** actinic rays
- **Syphilis** causes endarteritis obliterans and results in chronic superficial glossitis of the tongue which is a precancerous condition (rare these days).
- **Susceptibility**[1] of a person
- **Betel nut**, and slaked lime with betel leaf and tobacco (pan) is eaten and usually kept inside the cheek for many hours. Over the years, it brings about chronic irritation of mucosa of the cheek and causes leukoplakia. Tobacco contains multiple carcinogens including aromatic hydrocarbons.

Stages in the development of leukoplakia

I. **Keratosis** appears as a milky blush on the surface.
II. **Acanthosis** refers to elongation of rete pegs. This appears as a smooth, white, dry patch.
III. **Dyskeratosis** means the formation of keratin cell layer in the deeper aspect of epidermis, before they reach the surface.
IV. **Speckled leukoplakia** appears as multiple, small white patches on an erythematous base. ***It has the highest rate of malignant transformation*** (Figs 17.1 and 17.3).
V. **Carcinoma *in situ***

KEY BOX 17.3

CHRONIC HYPERPLASTIC CANDIDIASIS (Fig. 17.2)
Commissures of the mouth commonly affected
***A**lbicans Candida* invasion
No response to drugs, then surgery/laser treatment
Dense plaques of leukoplakia
Immunodeficiency can precipitate this condition
Dangerous because of malignant potential
Antifungal treatment—Topical application may help
Remember as **CANDIDA**

Fig. 17.1: Leukoplakia of the tongue—biopsy was positive for malignancy. He was treated with radiotherapy

Fig. 17.2: Chronic hyperplastic candidiasis affecting palate

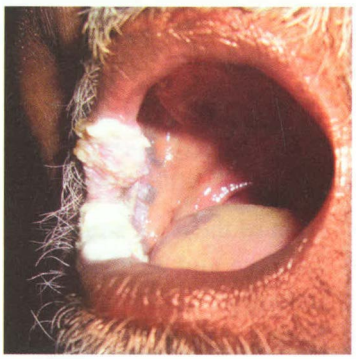


Fig. 17.3: Carcinoma arising from leukoplakia

(*Courtesy:* Dr Keerthilatha Pai, Head, Oral Medicine, College of Dental Sciences, Manipal)

[1]An 18-year-old girl, nonsmoker, nonalcoholic, with good oral hygiene had carcinoma tongue. What was the cause?

Treatment of leukoplakia

- About 10% of leukoplakia patients develop oral cancer. Hence, superficial excision of the lesion followed by skin grafting should be done.
- Even though leukoplakia is irreversible, **Isotretinoin** (13-cis-retinoic acid) can reverse some cases of leukoplakia and possibly reduce the development of squamous cell carcinoma.

2. **Erythroplakia** is a red, velvety lesion with an incidence of malignancy around 15% (17 times more malignant than leukoplakia). It is irregular in outline and may be nodular.
3. **Chronic hyperplastic candidiasis** (Key Box 17.3).
4. **Submucous fibrosis**
   - This is supposedly due to use of pan masala, arecanut with or without alcohol.
   - Initially it produces ulceration of mucosa of the cheek. These ulcers heal resulting in a dense submucous fibrosis, which appear clinically firm to hard. It can affect the tongue also. It is a progressive disease entirely confined to Asian population.
   - Chances of malignancy are around 10–15%.
   - Mouth opening may be restricted
   - It is treated by excision with reconstruction.
5. **Sideropenic dysphagia** (Plummer-Vinson and Paterson-Kelly syndrome). Iron deficiency occurs in the absence of anaemia in these patients. Common in Scandinavian women. Iron supplements reduce epithelial atrophy.
6. **Papilloma of the tongue or cheek**
7. **Discoid lupus erythematosis**
8. **Dyskeratosis congenita**
9. **Syphilitic glossitis:** Tertiary syphilis produces chronic superficial glossitis which can lead to carcinoma of the tongue. However, it is rare these days.
10. Human papilloma virus is a epitheliotropic virus. Its oncoproteins suppress tumour suppressor gene. It can give rise to tonsillar carcinoma and oropharyngeal carcinoma.
11. **Miscelllaneous** saw dust—Sinonasal adenocarcinoma.

- U-V rays—lip cancer
- Reverse cigarette smoking—palatal cancer.

Upper aerodigestive tract cancers

- Most of them are **squamous cell carcinomas.**
- **Tobacco and alcohol** are the most common aetiological factors.
- Most common premalignant lesion is **leukoplakia**.
- **Multiple anatomic sites** can be involved simultaneously (synchronous).
- **Second primary cancers** develop in 10–15% of cases **(metachronous).**
- Clinical presentation can be **peculiar/misleading** depending on anatomic site.
- **Generous biopsy** and MRI (if necessary) are the investigations of choice.
- Surgery, **radiotherapy and chemotherapy** are used singly or in combination in appropriate cases.

PEARLS OF WISDOM

Distant metastasis is more common with nasopharyngeal carcinoma than with any other head and neck cancer.

Common symptoms and sites (Table 17.1)

Table 17.1 Common symptoms and common sites of oral cancer (Ca = Carcinoma)

Symptom	Site of cancer—Ca
Pain around the eyes	Nasopharynx
Pain in the ear (otalgia)	Base of tongue, hypopharynx
Hoarseness	Glottis
Trismus	Extension of cancer into pterygoid muscles
Dysphagia	Base of tongue, hypopharynx, oesophagus
Loss of hearing	Auditory canal or nasopharynx

GENERAL PRINCIPLES IN THE TREATMENT OF ORAL CANCER

Aim of the treatment

1. **Cure of the patient:** Cure of the cancer, if possible, with wide excision of the tumour which includes removal of the tumour with 2 cm of the normal tissues, with or without bone.
2. **Palliation:** If cure is not possible, palliation should be attempted by surgery or radiotherapy.
3. **Preservation of function** such as swallowing, speech and vision, should also be taken into consideration.
4. **Cosmetic function:** Following wide excision, the cosmetic function must be maintained by reconstruction with myocutaneous/osteomyocutaneous flap.
5. To achieve **minimal mortality and morbidity**.
6. **Metastatic lymph nodes** are treated by neck dissection or curative radiotherapy (RT). Even when nodes are not palpable, following guidelines are given (Key Boxes 17.4 and 17.5).

KEY BOX 17.4

CLINICALLY 'NODE-NEGATIVE' NECK FROM ORAL CANCER

- Carcinoma lateral tongue, floor of mouth and mandibular alveolus commonly cause occult metastasis.
- Occult metastasis is seen in up to 30% of patients.
- Hence selective neck dissection of levels I, II and III are indicated in continuity with tumour excision in these cases.

KEY BOX 17.5

CLINICALLY NODE POSITIVE NECK

N1 : Selective supra-omohyoid neck dissection
{N2a N2b} : Modified radical or radical neck dissection followed by postoperative radiotherapy
N2c : Bilateral radical neck dissection—preserve at least one Internal Jugular Vein (IJV) + postoperative radiotherapy
N3 : Preoperative radiotherapy, if feasible radical neck dissection later.

TNM STAGING ORAL CANCER—American Joint Committee Cancer (AJCC)

PRIMARY TUMOUR (T)

- T0: No evidence of primary tumour
- Tis: Carcinoma *in situ*
- T1: ≤ 2 cm
- T2: > 2 cm and ≤ 4 cm
- T3: > 4 cm
- T4: Any cancer invading adjacent structures such as cartilage, cortical bone, deep (extrinsic) muscles of the tongue, skin or soft tissue of the neck.
  T4a: Moderately advanced local disease
  T4b: Very advanced local disease (skull base, pterygoid plate, internal carotid artery, masticator space)

REGIONAL LYMPH NODES (N)

- Nx: Nodes cannot be assessed
- N0: No lymph node metastasis
- N1: Single positive ipsilateral node less than or equal to 3 cm in greatest dimension
- N2a: Single positive ipsilateral node more than 3 cm but less than or equal to 6 cm
- N2b: Multiple ipsilateral nodes but all less than 6 cm
- N2c: Bilateral or contralateral lymph nodes but all less than 6 cm
- N3: Lymph node more than 6 cm.

DISTANT METASTASIS (M)

- M0: No distant metastasis
- M1: Distant metastasis present

STAGE GROUPING

Stage I	T1, N0, M0
Stage II	T2, N0, M0
Stage III	T3, N0, M0, T1–3, N1, M0
Stage IV	T4, N0, M0 , T, N2–3, M0
	T0, N0, M1

7. Treatment of advanced tumours—T3 and T4 lesions: These are managed by combination of surgery with postoperative RT. **Usually, surgery is the principal therapeutic modality of treatment followed by postoperative radiotherapy**. The treatment depends upon general condition of the patient, risks of anaesthesia, adequate intensive care management, etc. Chemotherapy also has been tried before or after surgery. However, response rate has improved but it has not affected the survival.

Flowchart showing treatment of primary tumour and metastasis is given in Figs 17.4 and 17.5

These are guidelines only, individualise the treatment.

Role of chemotherapy in head and neck cancers

- The most important benefit of chemotherapy has been in the treatment of laryngeal and nasopharyngeal carcinomas.
- **Cisplatin** is clearly the most effective drug. Other drugs such as carboplatin, 5-fluorouracil (5-FU), bleomycin, gemcitabine, etc. are also used.
- **Induction chemotherapy:** In advanced cases, chemotherapy is given before surgery or RT. In more than 80% of cases, tumour regression can occur.
- **Concurrent chemoradiotherapy** (CCRT): It improves both local and regional controls, specially in those patients with high risk cancers, e.g. locally advanced cancers of the oral cavity, larynx, oropharynx. Drug used in CCRT can be high dose cisplatin—100 mg/m^2 IV for 3 cycles every 21 days concomitantly with RT (for other dosage, kindly *refer* to oncology manual). Side effects include severe mucositis, xerostomia. Gastrostomy may be necessary for feeding.

Radiotherapy (RT) (Key Box 17.6)

- Irradiation of the oral cancers achieves a cure in about 80–90% of patients. It preserves anatomical part and also preserves the function.
- **The dose of RT:** 6500–7500 cGy units is required to eradicate squamous cell carcinoma of head and neck. It is usually given in the daily dose of 180–200 cGy units.

KEY BOX 17.6

RT: ADVANTAGES

- Easy, safe with minimal mortality
- Preservation of an organ
- Function of the part is preserved
- Cure rate is around 80 to 90%
- First line in early cases.

RT: DISADVANTAGES

- Long stay in the hospital (can also be taken as an out patient)
- Tumour cure cannot be assesed by pathology
- Soft tissue fibrosis resulting in ankylostomia
- Adverse effects on skin, loss of hair, mucositis of oral cavity, xerostomia, etc.

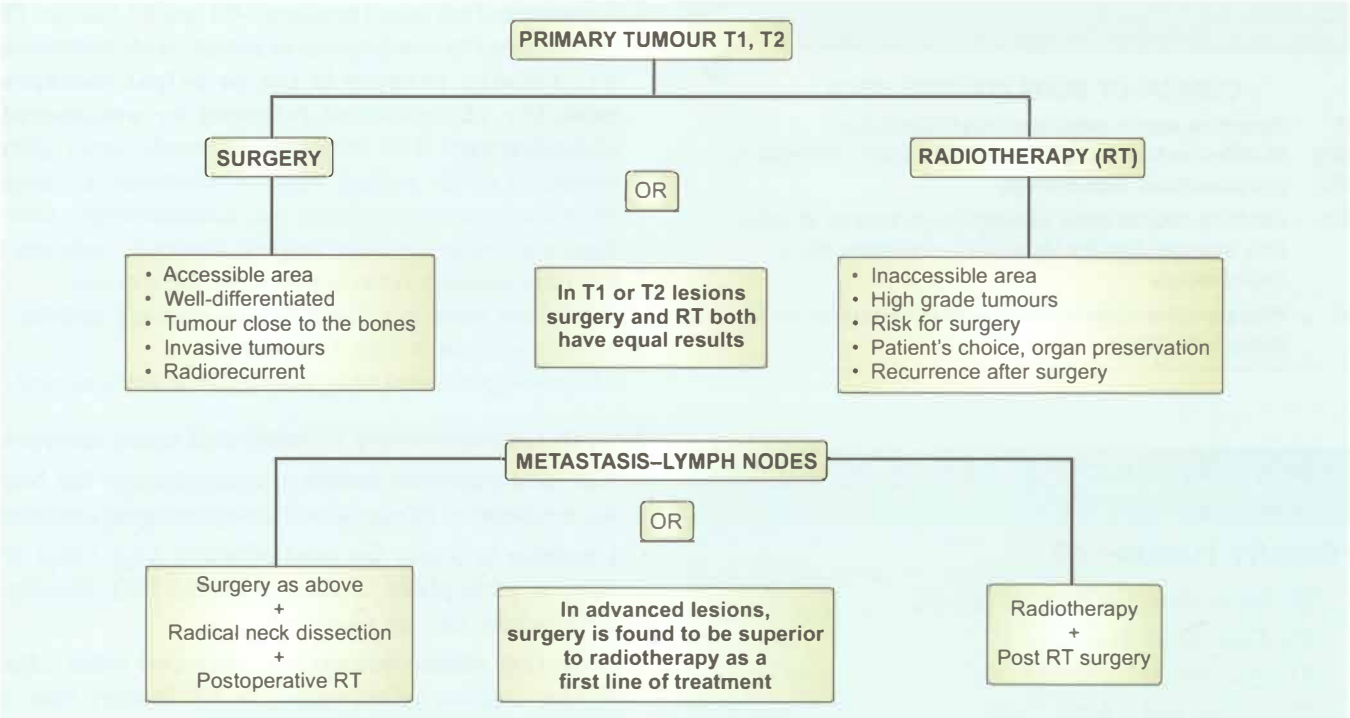


Fig. 17.4: Treatment of primary tumour and metastasis

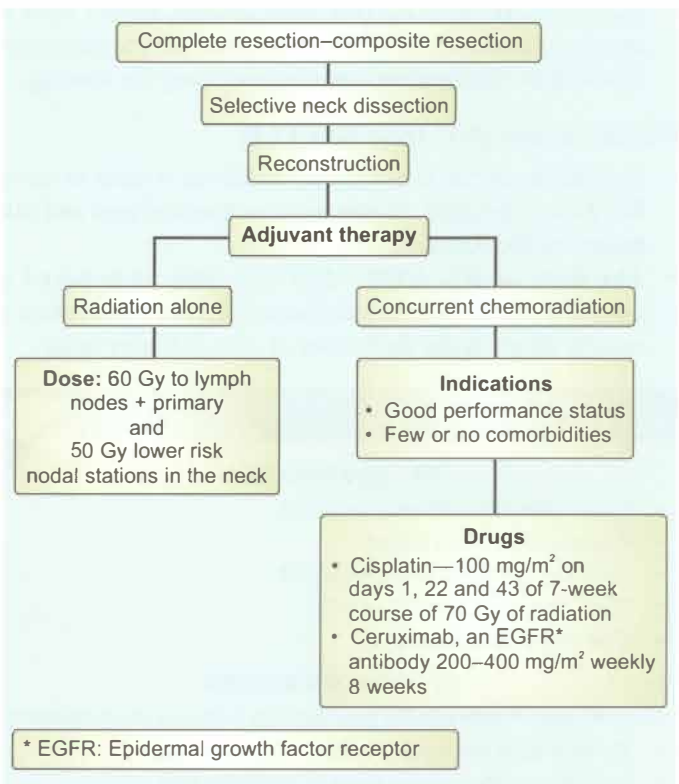


Fig. 17.5: Management of locally advanced nonmetastatic disease (III to IV B)

- Radiotherapy is given in T1 and T2 lesions as the first line of treatment and postoperatively in T3 and T4 lesions after surgery.

Role of surgery (Key Box 17.7)

- Surgery is done in all stages of oral cancers. It may be in the form of wide excision or **wide excision with removal of the bone (composite resection).** In advanced stages it may be palliative surgery such as excision of a fungating, ulcerating, bleeding mass. Surgery is also done for the lymph nodes in the form of radical neck dissection (RND), or modified RND.
- **The pectoralis major myocutaneous flap** (PMMC flap)**:** It is the most widely used flap for the reconstruction of oral cancers.

KEY BOX 17.7

SURGERY: ADVANTAGES

- It removes a fungating, ulcerating, bleeding lesion.
- It relieves the pain.
- The specimen is available for histopathological examination for cancer clearance.
- 80–90% cure is possible.

SURGERY: DISADVANTAGES

- Loss of an organ—total glossectomy
- Functional and cosmetic disability
- Significant morbidity
- Mortality: 8–10%.

Indications for surgery

- Early disease, bulky disease
- Tumour involving mandibular alveolus
- Tumour invading bone
- Advanced disease
- Fungating and bleeding lesion
- Radiorecurrence
- Multiple tumours
- Extensive premalignant changes of oropharyngeal mucosa

CARCINOMA OF BUCCAL MUCOSA

Carcinoma of the buccal mucosa is very common in India due to the habit of keeping **tobacco quid** in the cheek pouch (gingivoalveolar sulcus).

Pathological types

1. A **nonhealing** ulcer, with slough in the centre of the lesion
2. An **exophytic growth**, or a proliferative growth—verrucous carcinoma.
3. An **infiltrative lesion** slowly involves the adjacent structures such as tongue, mandible, floor of the mouth and skin. Skin infiltration results in orocutaneous fistula.

Clinical features

- A **nonhealing ulcer** or cauliflower like growth. Verrucous carcinoma is an exophytic growth.
- **Edges are everted** (Fig. 17.6) with induration at the base as well as at the edge. **Induration** clinically presents as a hard feeling. Pathologically, it is due to fibrosis, caused by malignancy (carcinomatous fibrosis). It is a diagnostic feature of squamous cell carcinoma. Possibly, it is a host reaction indicating good immunity. Due to fibrosis, some lymphatics get obliterated. This delays spread of the disease, thereby improving the prognosis.
- Proliferative lesions are often verrucous carcinoma (Key Box 17.8).
- Ulcer **bleeds on touch**. Due to secondary infection most of the oral cancers are tender to touch (Fig. 17.9).

KEY BOX 17.8

PECULIARITIES OF VERRUCOUS CARCINOMA

- Very slow-growing
- Growth is exophytic (than infiltrative)
- Rarely spreads by lymphatics
- It is a well-differentiated carcinoma
- Surgery is the treatment of choice.

- **Fixity** to the underlying structures such as mandible may be present.
- **Surrounding area** may also show **induration**.
- Evidence of **leukoplakia** may be present in the oral cavity.
- **Trismus** is due to involvement of pterygoid muscles and masseter. This occurs when carcinoma buccal mucosa **extends into the retromolar trigone**. Trismus can also be due to soft tissue fibrosis caused by radiation. **Once perineural lymphatics are involved, spread can occur in infratemporal fossa resulting in trismus** (Key Box 17.9 and Fig. 17.10).
- **Halitosis** is very characteristic.

KEY BOX 17.9

TRISMUS

Difficulty in opening the mouth is called trismus. Normal mouth opening ranges from 35 to 45 mm.

Grades:

- Grade I : Mouth opening is between 2.5 and 4 cm
- Grade II : Between 1 and 2.5 cm
- Grade III: Less than 1 cm

Common causes of trismus

1. Temporomandibular joint involvement—such as ankylosis, dislocation, synovitis, etc.
2. Mandible fractures
3. Pterygoid muscle infiltration by growth in the retromolar region
4. Acute inflammatory lesions in the oral cavity
5. Tetanus and tetany
6. Radiation fibrosis of soft tissues/muscles of mastication

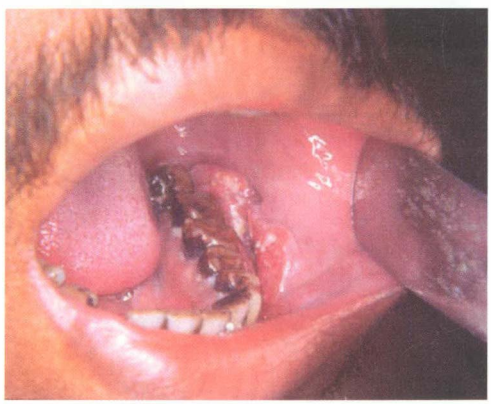


Fig. 17.6: Carcinoma alveolar margin. Look at teeth stains—everted edge

Fig. 17.7: Carcinoma cheek with orocutaneous fistula

Fig. 17.8: Carcinoma buccal mucosa infiltrating skin—'warning' of fistula

- Assessment of fixity to mandible: Severe pain over the jaw indicates periostitis.
- **Bidigital palpation of mandible** is done by examining with index finger on the outer aspect of the mandible and the thumb on the under surface of the mandible. This test should be done on the opposite side first. Only then, the thickening of the mandible can be appreciated.
- **Gingival cancers** (Fig. 17.11)
  - Early cases present as mucosal change in leukoplakia
  - Loosening of tooth may be a presenting feature.
  - Can present as bleeding and pain
  - Bone involvement occurs early
  - Spread to adjacent structures occurs early.

Spread

1. **Local spread:** Once it involves the entire thickness of the cheek it results in orocutaneous fistula (Figs 17.7 and 17.8). Involvement of mandible results in sinus (Key Box 17.10).

KEY BOX 17.10

CARCINOMA BUCCAL MUCOSA AND MANDIBLE

- Direct infiltration by the tumour
- Through mandibular canal
- Through periodontal membrane
- Orthopantomogram or spiral CT can be used for imaging
- Loss of central part of mandible results in pouting of lower lip and continuous drooling of saliva. It is called as **Andy Gump deformity**[1].

2. **Lymphatic spread:** Submandibular nodes and upper deep cervical nodes get enlarged (Levels I and II). In 50% of the cases, lymph node enlargement is due to infection and remaining 50%, it is due to metastasis. Metastatic deposits are hard in consistency, indurated and with or without fixity. Significant oedema of face can occur due t lymphatic spread.
3. **Blood spread:** It is very rare and it occurs late.

PEARLS OF WISDOM

The mandibular canal is close to occlusive surface, in edentulous elderly patients due to decrease in the vertical height of horizontal ramus thus facilitating easy spread (Figs 17.12 and 17.13) ***of oral cancer to mandible.***

Investigations

1. **Wedge biopsy from the edge of the ulcer** is taken because of the following reasons:
   - Tumour cells are concentrated more in the growing edge
   - Comparison with the normal tissues is possible.
   - Centre of the ulcer has slough.
   - **Histopathological report** shows squamous cell carcinoma and in majority of the cases it is well-differentiated.
2. **Orthopantomography:** X-ray of mandible to rule out mandibular involvement
3. **Chest X-ray** to detect inhalation pneumonia.
4. **FNAC** of the lymph node.
5. **Magnetic resonance imaging (MRI)**
   - Large advanced lesion can be better assessed by MRI. Soft tissue infiltration can be assessed correctly thus dictating the extent of resection especially in patients with restricted mouth opening.
   - **It is the investigation of choice to look for involvement of skull base, brachial plexus, spinal nerve roots and lymph nodes.**
   - MRI has no radiation hazards.

Treatment of carcinoma buccal mucosa

It can be classified into early disease and advanced disease (Figs 17.9 to 17.11).

Fig. 17.9: Carcinoma buccal mucosa — advanced

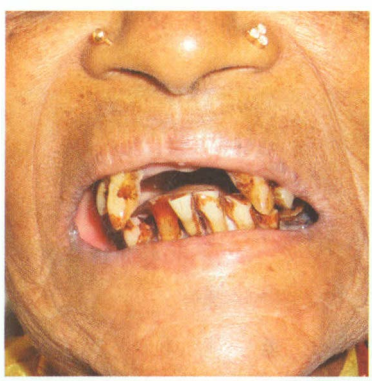


Fig. 17.10: Carcinoma buccal mucosa infiltrating mandible — severe trismus. Also observe tobacco stains

Fig. 17.11: Carcinoma alveolus

[1]**Andy Gump** was one of the characters in a popular comic strip, 'The Gumps' created by Sidney Smith in 1917. The character's face seems to end at the upper lip due to 'absence of mandible' and is chinless. A statue of this comic character is on display at Lake Geneva Museum

Early disease

Two modalities are given in Key Box 17.11.

KEY BOX 17.11

EARLY CARCINOMA BUCCAL MUCOSA

- T1, T2 lesions—surgery/RT
- T1 lesion near commissure—RT
- T2—exophytic and superficial—RT
- T2—deep—surgery is better
- Early disease—no nodes—surgery is better—no other treatment is necessary.
- Early disease—positive lymph nodes—same modality to be used for primary and secondary.

I. Surgery

1. A small **superficial ulcer** (T1, T2) is treated by wide excision followed by split skin graft (SSG).
2. An **infiltrative lesion** is treated with wide excision followed by a flap reconstruction. Usually, PMMC (pectoralis major myocutaneous flap) is used.

   PMMC flap: This is the most widely used flap now for head and neck reconstruction. The flap is raised along with muscle and an island of skin based on pectoral branch of thoracoacromial artery. It is tunnelled under the skin of chest wall and neck and brought to the area of the defect. It has been described as the **'workhorse'** for head and neck reconstruction.

- **Radial artery based flap** is the **workhorse of microvascular reconstruction.**

II. Radiotherapy

As mentioned earlier, early lesions can be managed with radiotherapy (RT). The advantage of RT is that it cannot only cure the disease but also preserve the organ and its function.

Indications for radiotherapy

1. Patient not willing for surgery
2. Patient not fit for surgery
3. T1 and T2 lesions
4. Lesion near the commissure.

Types

1. **External radiotherapy:** Large total dose of 6000–8000 cGy units are given at the rate of 200 cGy units/day.
2. **Interstitial radiotherapy** is indicated in infiltrative small lesions. Caesium 137 or iridium wires are placed within the tumour. Advantage of this method is minimal tissue reaction.

Advanced carcinoma buccal mucosa

(Figs 17.14 to 17.19)

Surgery: T3 and T4 lesions require surgery as the main modality of the treatment followed by postoperative radiotherapy. Most of the lesions require full thickness resection leaving behind large defects. Such defects can be repaired using myocutaneous flap.

PEARLS OF WISDOM

When the primary lesion is removed en bloc with mandible and cervical lymph nodes, it is called composite resection.

Examples of surgeries

1. **Carcinoma buccal mucosa fixed to the mandible:** Wide excision of the growth along with segmental resection of the mandible or hemimandibulectomy is done depending upon the infiltration of the tumour. Very often, whole thickness of the cheek is lost which is reconstructed by using PMMC flap (*see* next page).

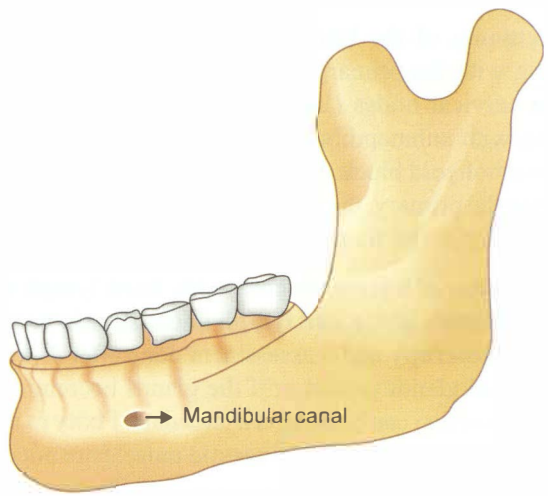


Fig. 17.12: Mandibular canal in a normal person is well away from occlusive surface

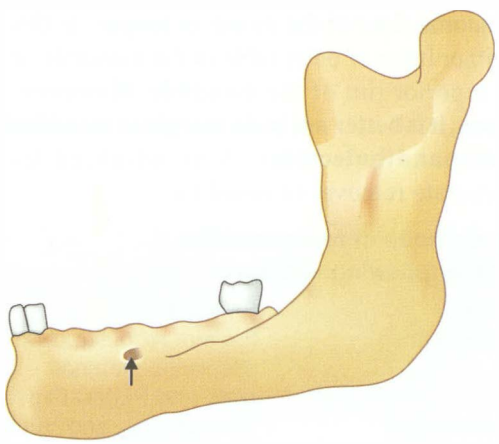


Fig. 17.13: Mandibular canal in an edentulous patient is close to occlusive surface explaining easy spread in carcinoma buccal mucosa

Fig. 17.14: Carcinoma cheek with infiltration into the skin and mandible—locally advanced. Good case for **composite resection**

Fig. 17.15: Neglected carcinoma buccal mucosa infiltrating floor of the mouth, commissure and lower lip

Fig. 17.16: Advanced carcinoma buccal mucosa with involvement of half of lower lip

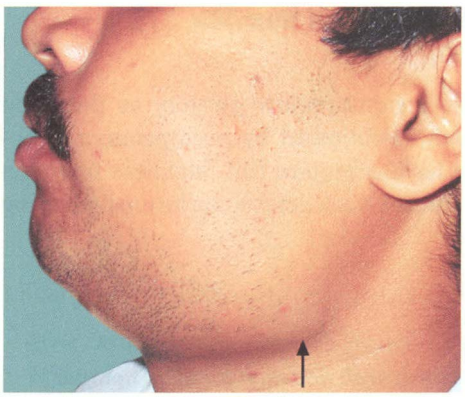


Fig. 17.17: Gross lymphoedema of left side of the face with enlargement of submandibular lymph nodes

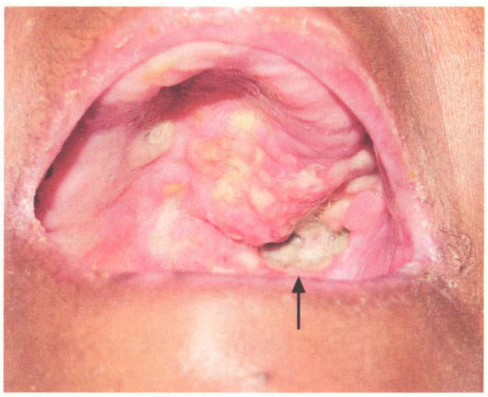


Fig. 17.18: Carcinoma alveolus—excavating ulcer with slough, infiltrating retromolar trigone. RT followed by surgery is the ideal choice of treatment

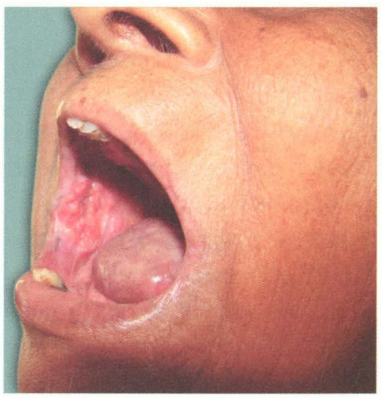


Fig. 17.19: Carcinoma buccal mucosa on the right side arising from submucous fibrosis

- **Segmental mandibulectomy:** *Indications*
  A. Clinical and radiological involvement of bone
  B. To obtain wide margins
  C. To facilitate reconstruction when one has to use 'bulky' PMMC flap
  D. Excision of full thickness of cheek.
- **Marginal mandibulectomy:** It can be done for carcinoma floor of the mouth or tongue. In this removal of either inner or outer table of the mandible or excising the superior rim of the mandible. **However, in large lesions, it is better not to do marginal mandibulectomy.**
- **Hemimandibulectomy:** Very advanced lesion may necessitate removal of mandible.

Various methods of reconstruction of mandible following excision

• Soft tissue	: PMMC flap
• Soft tissue with bone	: 2.4 mm reconstruction alloplastic material plate and PMMC flap
• Nonvascularised bone grafts	: Titanium tray and cancellous chips from iliac crest
• Vascularised bone grafts	: Fibula, iliac crest

2. **Orocutaneous fistula** is treated by wide excision which refers to removal of the entire thickness of the cheek along with the growth.

   Reconstruction is done by using PMMC flap. Radiotherapy should not be given as it results in persistence of fistula.

3. **Carcinoma of the buccal mucosa with lymph nodes:** Along with the primary, submandibular nodes and upper deep cervical nodes (Levels I, II and III) are removed, along with submandibular salivary gland. This is called supraomohyoid block dissection. If surgery has been used to treat the primary, the lymph nodes also should be treated by surgery in the form of neck dissection.

4. **Carcinoma of buccal mucosa with fixed lymph nodes:** **Both primary lesion and lymph nodes should be treated by radiotherapy** and reassessment done after 3–4 weeks. If residual glands persist or if the glands become mobile, neck dissection can be done at a later date. Fixity to internal jugular vein or sternocleidomastoid muscle are not contraindications for radical block dissection. Those structures can be removed. However, when the lymph nodes are fixed to the carotid artery, radiotherapy is preferred.

Prophylactic neck dissection

- It is advocated in T3 and T4 lesions irrespective of nodal status. This amounts to minimal supraomohyoid neck dissection with removal of Levels I, II and III lymph nodes. It has shown survival benefits.
- It is also indicated in carcinoma mandibular alveolus or buccal mucosa extending into the floor of mouth.

Complications of carcinoma buccal mucosa

1. Orocutaneous fistula
2. Trismus—It can be due to direct infiltration of pterygoid muscles or masseter or soft tissue fibrosis following RT.
3. Recurrent respiratory tract infection
4. Cancer cachexia.

STEPS OF PECTORALIS MAJOR MYOCUTANEOUS FLAP (PMMC) (Figs 17.20 to 17.25)
WORKHORSE OF HEAD AND NECK RECONSTRUCTION

Fig. 17.20: Amount of tissue to be removed (wide excision) is marked. Horizontal incision is given in the neck for block dissection

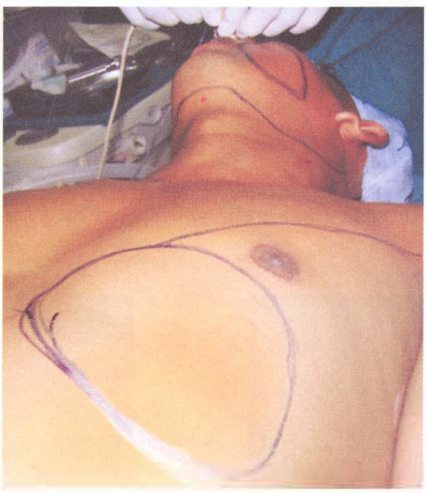


Fig. 17.21: Skin paddle outlined prior to elevation

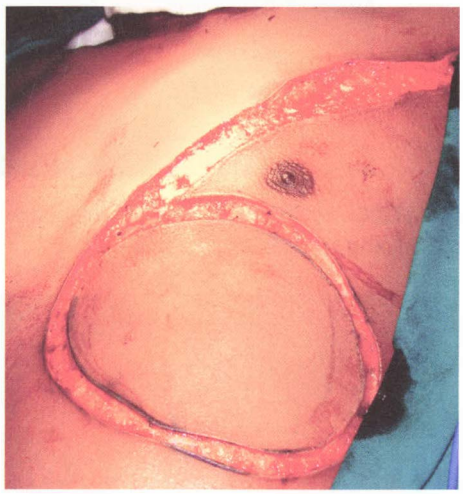


Fig. 17.22: Skin paddle mobilised

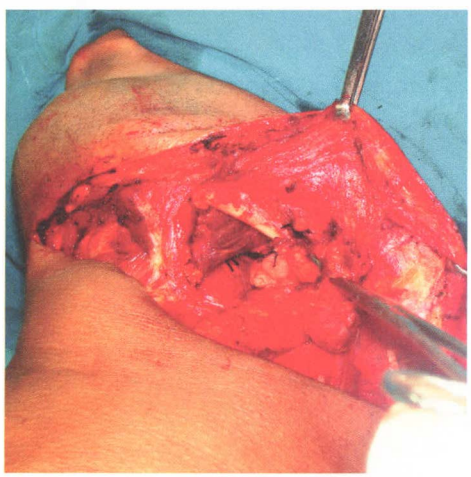


Fig. 17.23: Elevation of flap

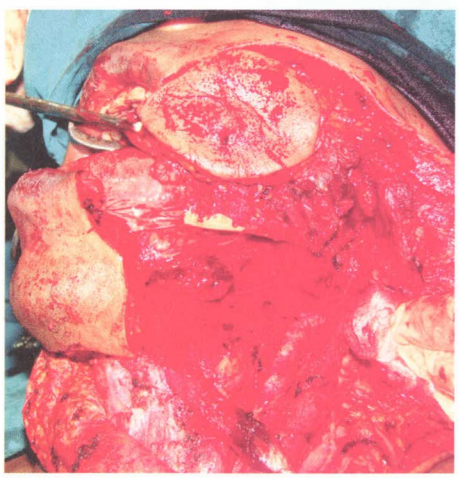


Fig. 17.24: You can see here wide excision of growth of mandible. The specimen is about to be removed

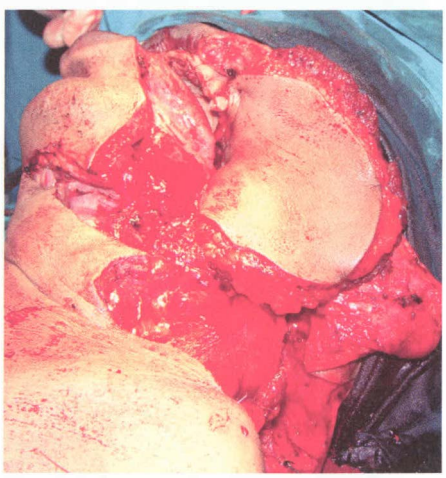


Fig. 17.25: Elevated flap is being mobilised

(*Courtesy*: Prof. Satadru Ray, Head of the Department, Surgical Oncology, KMC, Manipal)

PMMC FLAP—CONTINUED (Figs 17.26 to 17.31)

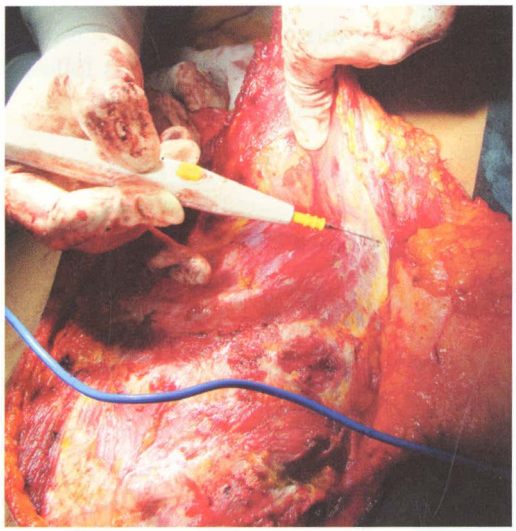


Fig. 17.26: Flap being turned out (bipedaled) to provide inner and outer lining for the tissue loss

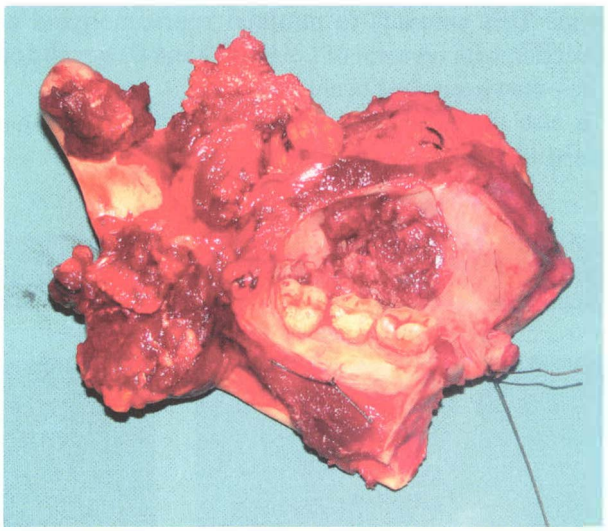


Fig. 17.27: Composite resection: Inner aspect

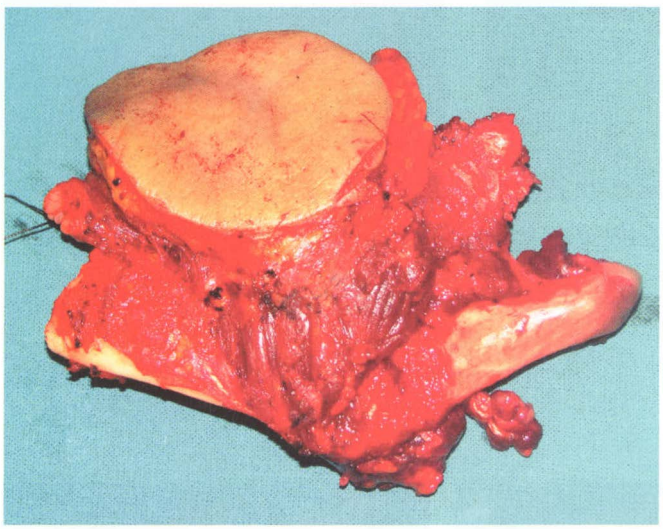


Fig. 17.28: Composite resection: Outer aspect

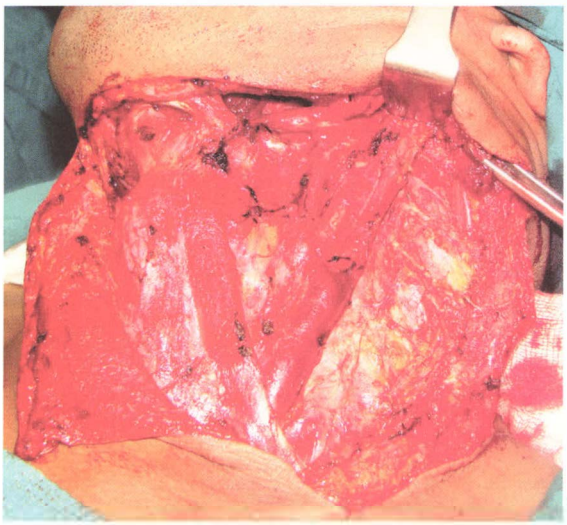


Fig. 17.29: Modified radical neck dissection is being done

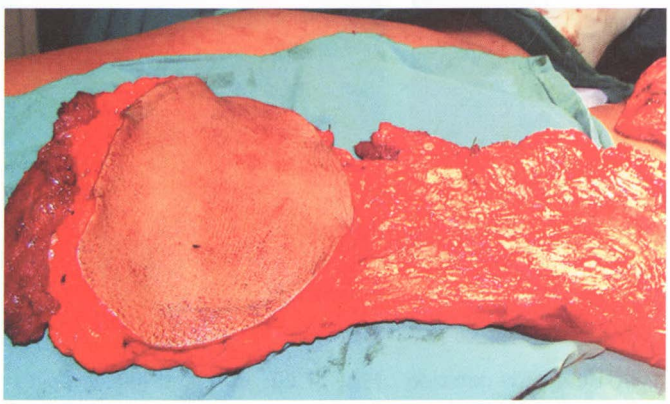


Fig. 17.30: Completely mobilised PMMC flap ready for reconstruction. Main vessels in the paddle are **pectoral branches of acromiothoracic artery.** Mandible reconstruction is not mandatory

Fig. 17.31: Healing after 10 days

CARCINOMA OF TONGUE

Pathological types

1. Nonhealing ulcer, commonly on lateral border of tongue in 60% of cases, with slough (Fig. 17.32)
2. A proliferative growth, with everted edge
3. Frozen tongue or indurated variety (Fig. 17.33)
   In this variety, there is maximum induration and sometimes it is more than the size of tumour. The tongue is converted into a hard woody "mass".
4. ***Fissure variety:*** The tongue is indurated with deep fissure.

Clinical presentation

- A bleeding ulcer
- Pain in the tongue is due to involvement of lingual nerve. In such cases, pain from the tongue can be referred to the ear and lower temporal region.[1]
- **Ankyloglossia** is restricted mobility of the tongue. It is due to infiltration of the floor of the mouth or mandible, or due to an advanced lesion (Figs 17.33 and 17.34).
- **Disarticulation**—difficulty in talking is due to inability of the tongue to move freely.
- **Dysphagia** is a common presentation from carcinoma of posterior 1/3rd (in 20% cases). An elderly gentleman sitting in the outpatient department spitting blood-stained saliva is suggestive of carcinoma posterior 1/3rd of the tongue (Figs 17.35 and 17.36).
- **Foetor oris** is due to infected necrotic growth.

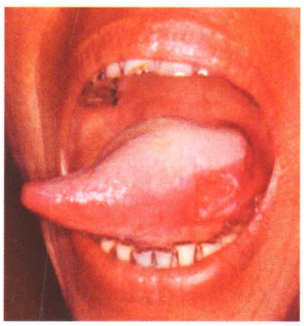


Fig. 17.32: Carcinoma tongue lateral border—the most common site

Fig. 17.33: Frozen tongue—ankyloglossia and dysphagia are present

Fig. 17.36: Carcinoma tongue with absolute dysphagia, fixed lymph nodes in the neck and involvement of mediastinal lymph nodes-receiving radiotherapy—on Ryle's tube feeding

Fig. 17.34: Carcinoma tongue with ankyloglossia, disarticulation, dysphagia

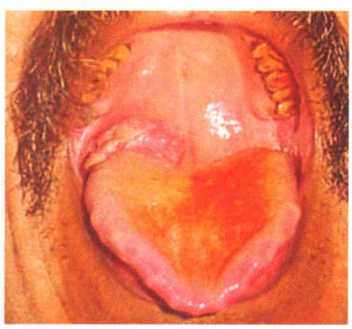


Fig. 17.35: Carcinoma posterior 1/3rd easily missed

Fig. 17.37: Carcinoma tongue—excavating ulcer

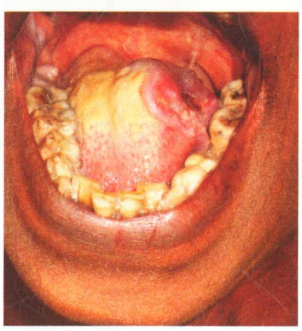


Fig. 17.38: Carcinoma tongue left lateral border everted edges and excavating

Fig. 17.39: Carcinoma lateral border tongue in a 40-year-old man—elevated and everted lesion

[1]Auriculotemporal nerve and lingual nerve are posterior branches of mandibular division of trigeminal nerve.

- **Bilateral massive enlargement** of lower deep cervical nodes in an elderly patient is suggestive of carcinoma of posterior 1/3rd. The patient may not be aware of growth at all.

PEARLS OF WISDOM

Tongue cancers tend to be more rapid in their onset than other cancers in the oral cavity. Compared with other cancers within the oral cavity, tongue cancers have greater potential of lymph node metastasis.

Clinical examination

- Inspection and palpation of the growth or the ulcer should be described in the same manner as that of carcinoma cheek. Typically, the ulcer bleeds on touch with central slough. The edge, base and surrounding area are indurated. Carcinoma of the tongue and carcinoma of the penis are two places in the body wherein induration can be much more extensive than the primary growth or an ulcer. In some cases, induration may be the only finding. Everted edge is commonly seen (Figs 17.37 to 17.39).
- Digital palpation of posterior 1/3rd of tongue should be done with a glove.
- **Test for mobility of the tongue.**
  - **Forward** protrusion—**genioglossus**. This is the muscle commonly involved.
  - **Backward** movement—**styloglossus**
  - **Elevation—palatoglossus**
  - **Depression—hyoglossus**

  All these muscles are supplied by hypoglossal nerve except palatoglossus which is supplied by glosso-pharyngeal nerve.
- **Bidigital palpation** of the mandible should be done which may show thickening.

Lymphatic spread (Fig. 17.40)

1. **Apical vessels** drain the tip of the tongue into submental lymph nodes, bilaterally.
2. **Lateral vessels** drain into submandibular lymph nodes, from here to the lower deep cervical lymph nodes and jugulo-omohyoid nodes—level III.
3. **Central vessels** drain into submandibular nodes.
4. **Basal vessels** drain the posterior 1/3rd of the tongue. There is criss-crossing of the lymphatics on both sides. Hence, they drain into bilateral lower deep cervical lymph nodes. (Some lymphatics on their way to the nodes, pass through the mandible which explains the frequent involvement of mandible in carcinoma tongue. This is a debatable issue now.)
   - In 50% of cases, the lymph node enlargement is due to secondary infection. Such nodes are tender and firm and respond to antibiotics. In remaining cases, they are hard and fixed and hence, significant.

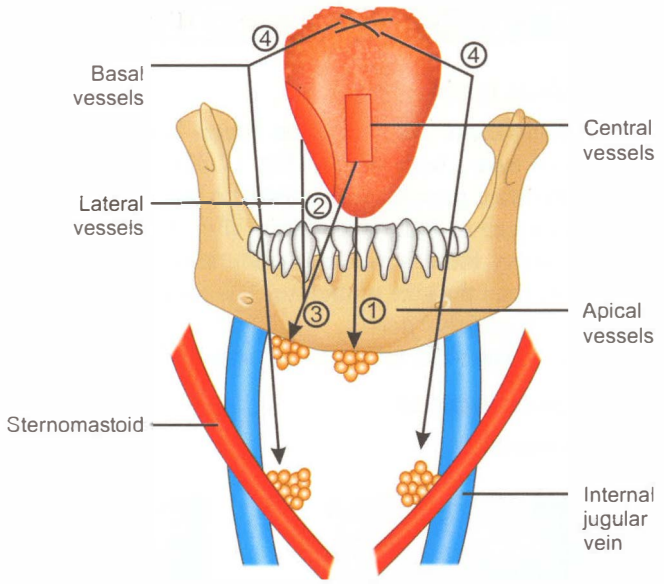


Fig. 17.40: Lymphatic drainage of the tongue—*see* text for numbers

PEARLS OF WISDOM

Posterior 1/3 tongue has very less cornification but has abundant lymphatics which explains massive nodes (Key Box 17.12).

Investigations

1. **Wedge biopsy from edge of the ulcer** can be taken under local anaesthesia. In cases of **proliferative growth, punch biopsy is recommended.** In cases of growth arising from posterior 1/3rd of the tongue, biopsy can be taken under general anaesthesia. It also provides an opportunity to examine in detail the posterior spread of the disease into tonsils, pharynx, etc.
2. **Orthopantomogram:** X-ray of the mandible can demonstrate an irregular defect due to **invasion, erosion** or **pathological fracture.**

KEY BOX 17.12

CARCINOMA POSTERIOR 1/3rd

- It presents with **dysphagia** or with a change in voice.
- **Easily missed** in a clinical examination
- **Biopsy** should be done under **general anaesthesia** to avoid aspiration and to assess the spread posteriorly.
- Palpation will give the diagnosis—induration
- It is one of the **occult primaries** for lymph node secondaries in the neck.
- Criss-crossing of the lymphatics explain **bilateral lymph** nodes in the neck.
- **Blood spread** is more common.
- Prognosis is **bad** because well-differentiated carcinoma in this location is rare.

3. **Chest X-ray** is taken to rule out **aspiration** or **inhalation pneumonia.**
4. **Routine investigations** such as complete blood picture, fasting and postprandial sugar estimation to rule out diabetes and electrocardiography to assess cardiovascular function should be done.
5. **Very advanced lesions** may require CT scan/MRI scan.

Treatment

Carcinoma of the tongue is managed similar to a cancer in the oral cavity. However, to preserve the function of the tongue, widespread disease in the posterior one-third tumours, general health of patient (elderly with bad bronchopneumonia) may decide the treatment in favour of radiotherapy. However, results of surgery or radiotherapy for early carcinoma of tongue are equivalent.

Various types of surgery

1. **Carcinoma *in situ*:** This type is uncommon in our country. Wide excision with 1 cm margin and a depth of 1 cm is sufficient. Reconstruction of the tongue is not necessary.
2. **Partial glossectomy** is indicated when the lesion is less than 2 cm (T1) and confined to the lateral border of the tongue. About 1/3rd of the anterior 2/3rd of the tongue is removed. The wide excision should include at least 2 cm of tissue away from the palpable indurated edge of the tumour (Figs 17.41 and 17.43).
   - Alternatively, radiotherapy can be given.
3. **Hemiglossectomy** refers to removal of around 50% of the tongue. This is indicated in a radio-residual tumour, radio-recurrent tumour or where radiotherapy facilities are not available (Fig. 17.42). Reconstruction of the tongue can be done by nasolabial flap and division of pedical at later date. Radial forearm free flap can also be used.
4. **Total glossectomy:** Indications are similar to those mentioned above. However, very extensive growth involving the entire tongue are given radiotherapy initially, to reduce the size of the tumour. Surgery can then be undertaken. Total glossectomy carries significant mortality and morbidity.

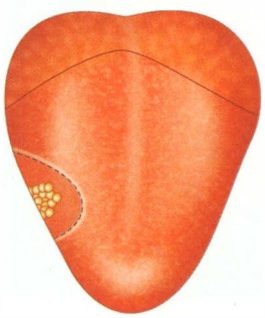


Fig.17.41: Partial glossectomy

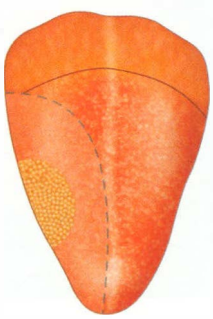


Fig. 17.42: Hemiglossectomy

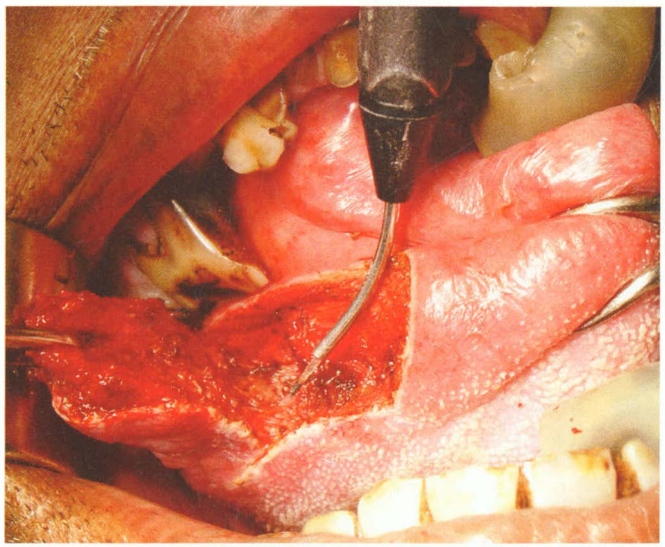


Fig.17.43: Partial glossectomy done with the help of laser—excellent tool for haemostasis (*Courtesy:* Dr Balakrishnan, Prof of ENT and Head and Neck, KMC, Manipal)

5. **Commando's operation:** This is indicated when carcinoma of tongue is fixed to the mandible with infiltration of the floor of the mouth. Hemiglossectomy with hemimandibulectomy, removal of the floor of the mouth and radical neck dissection is described as Commando's operation (Key Box 17.13).
   - However, in a few selected cases, removal of the **haemimandible is not necessary**. Growth which is close to the margin of the mandible without infiltration (confirmed by X-ray) needs to be treated by **marginal mandibulectomy**. Carcinoma of the tongue with involvement of only a small portion of mandible can be managed by **segmental excision**. Advantage of this method is that it is not only cosmetic but also **preserves the function of the tongue by preserving genio-glossus**. Hence, the tongue may not fall backwards after surgery.

KEY BOX 17.13

STRUCTURES REMOVED IN RADICAL BLOCK DISSECTION OF THE NECK

- The fat, fascia, lymphatics from midline to the anterior border of trapezius, from mandible to clavicle below.
- The lymph nodes—submental, submandibular, upper and lower deep cervical nodes, posterior group of nodes (Levels I–V).
- Submandibular salivary gland, sternomastoid, one side internal jugular vein (IJV) are sacrificed
- Spinal accessory nerve is removed
- Lower pole of parotid is removed to facilitate lymph node clearance.

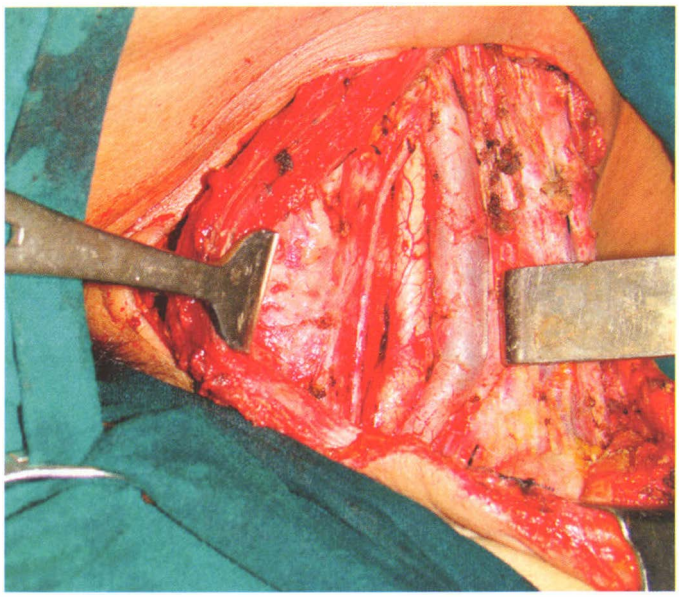


Fig.17.44: Modified radical neck dissection is in progress—internal jugular vein, carotid artery and vagus nerve are seen

TREATMENT OF LYMPH NODES (Fig. 17.44)

- Lymph node metastasis in the neck from squamous cell carcinoma can be managed both by surgery as well as radiotherapy. Radiotherapy can be given in all stages of secondaries in the neck. However, its main indication is a large primary tumour with neck nodes. In such situations both the primary and secondary can be managed with radiotherapy alone which carries minimal morbidity and mortality.
- If the general condition of the patient is good and the lymph nodes are hard and mobile, hemiglossectomy with excision of the floor of the mouth with radical dissection of the neck is done (Commando's operation).
- **If radical neck dissection has to be done on both sides, the IJV should be preserved at least on one side to prevent cerebral oedema. In such cases, radiotherapy is a very good alternative.**

Please note: Details about radical neck dissection is given in Chapter 16, page 267.

Causes of death in carcinoma tongue

1. Recurrent aspirational pneumonia
2. Gross local recurrence, fungation, ulceration, cachexia.
3. **Uncontrolled haemorrhage from growth:** In such cases ligation of external carotid artery above superior thyroid branch should be done (Fig. 17.45). If ligature is applied below the origin of superior thyroid artery, it results in eddy currents and thrombus at bifurcation of common carotid artery.

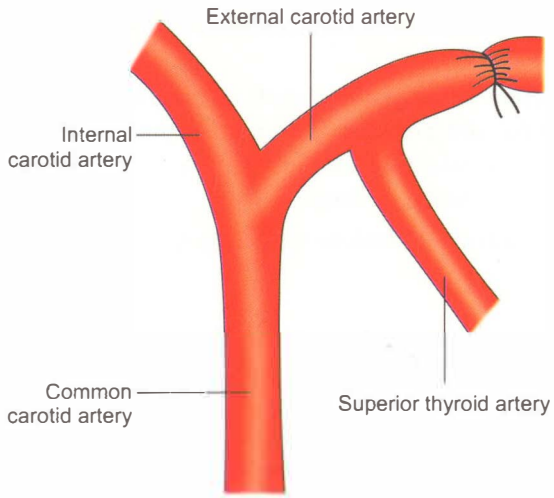


Fig. 17.45: Ligation of external carotid artery

CARCINOMA OF LIP

- Incidence of carcinoma of the lip is about 10 to 12%. It is common in the western, elderly, white people, specially those exposed to sunlight. The actinic rays produces actinic cheilitis—inflammation of the lip, especially lower lip, which over a period of years can turn into malignancy.
- Since this is common in agriculturists, who are constantly exposed to sunlight, it is called **Countryman's lip** (Key Box 17.14).

KEY BOX 17.14

COUNTRYMAN'S LIP

Sunlight
↓
Actinic rays
↓
Cheilitis
↓
Erythema
↓
Cracks

Fig. 17.46: Advanced carcinoma lip—a proliferative lesion, treated with radiation

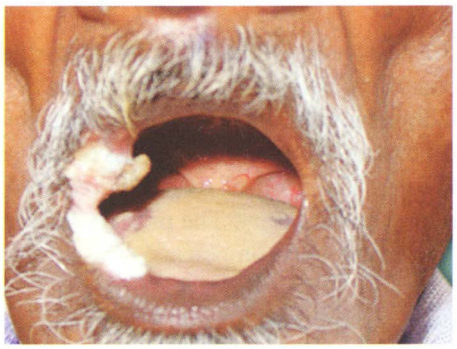


Fig. 17.47: Carcinoma lip—everted edge and exophytic growth—also observe coating of the tongue

Fig. 17.48: Carcinoma lip with infiltration of buccal mucosa. Requires lip and cheek reconstruction

Fig. 17.49: A bleeding exophytic lesion. Better managed by surgery than radiotherapy

- Carcinoma lip includes growth arising from vermilion surfaces and mucosa.
- **Leukoplakia** is also responsible for squamous cell carcinoma. Smoking, spirits and spices are the common precipitating factors.
- **Genetic factors** also may play a role. Blacks are less susceptible. On the other hand, **increased incidence** of carcinoma lip has been found in **Caucasians.**
- **Khaini chewers** are more susceptible for carcinoma of the lip (*khaini* is a mixture of tobacco and lime).
- It can also present as verrucous carcinoma of lip.

Clinical features (Figs 17.46 to 17.50)

- Elderly males are affected in 90% of cases.
- **Nonhealing** ulcer or growth is a common presentation.
- **Edge is everted and indurated.** Induration of the edge and the base is characteristic (Fig. 17.47).
- Floor is covered with slough. Bleeding spots may be visible.
- **Mobility:** Ulcer or the growth moves with the lip, it is fixed to the subcutaneous structures of the lip.
- The entire upper lip and lateral portions of the lower lip drain into **upper deep cervical nodes.** Central portion of the lower lip drains to submental nodes and submandibular nodes. Like elsewhere in the oral cavity, in 50% of the cases, nodes are enlarged due to secondary infections. In remaining 50% of the cases, they are enlarged due to metastasis. Such nodes are hard, with or without fixity. Blood spread is uncommon.

Fig. 17.50: Very large lesion started in the upper lip—later involving the lower lip also—case of kiss cancer

Differential diagnosis (Key Box 17.15)

KEY BOX 17.15

DIFFERENTIAL DIAGNOSIS OF CARCINOMA LIP

- Keratoacanthoma
- Ectopic salivary gland tumour
- Pyogenic granuloma
- Leukoplakia.

In a classical case of carcinoma of the lip with everted edges and induration, there is no differential diagnosis. However, following are a few conditions to be remembered:

1. **Keratoacanthoma**
   - It is a cutaneous tumour arising from hair follicles on the lips. It is common in white, western, males between 50 and 70 years of age.
   - Sunlight (actinic rays), chemical carcinogen, viral factors may be responsible for this lesion.
   - The central portion of the nodule may ulcerate. The lesion may progress for 6 weeks and may resolve spontaneously within 4–6 months.
2. **Ectopic salivary gland tumour**
   - The lip is one of the common sites of malignant salivary gland tumours. This presents with submucous nodules that grow slowly and ulcerate and may mimic squamous cell carcinoma (Fig.17.51).
   - They are also indurated lesions.
   - However, the characteristic everted edge may not be seen.
   - These are adenocarcinomas which are treated by surgery.
3. **Pyogenic granuloma** (Fig. 17.52)
   - Recurrent infections or trauma produces a polypoidal mass with significant bleeding.
   - It is rich in granulation tissue and resembles a polyp.
   - It is devoid of epithelium
   - Histologically, it is a capillary haemangioma
   - Absence of induration gives the diagnosis.

Fig. 17.51: This lesion was diagnosed as carcinoma lip. However, it did not have everted edges. It was indurated. Biopsy reported as ectopic salivary gland tumour. On careful questioning, patient says it started as a swelling not as an ulcer

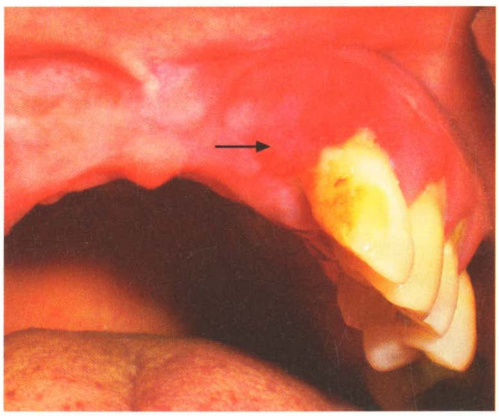


Fig. 17.52: Pyogenic granuloma due to caries tooth (*Courtesy:* Dr Keerthilatha Pai, HOD, Department of Oral Medicine, College of Dental Sciences, Manipal)

4. Leukoplakia

A slow developing leukoplakia presents as whitish nodule or an ulcer. However, biopsy confirms the diagnosis.

Treatment

Surgery and radiotherapy are the two modalities available for the treatment of carcinoma of the lip.

I. Surgery

- T1 and T2 lesions can be excised followed by direct suturing without much functional problems. This is described as "V" excision which includes removal of growth with 1 cm healthy margin. Care should be taken to excise full thickness of the lip.
- When removal of more than 1/3rd of the lip is required, flap reconstruction may be necessary. The primary goal in lip reconstruction surgery is oral competence.

  Examples

  1. **Abbe flap:** Based on upper labial artery—a pedicled flap is rotated down and sutured to the defect at the lower lip (Fig. 17.55).
  2. **Estlander's flap:** Wedge-shaped flap is used to reconstruct carcinoma of lower lip, when it involves the angle (Fig. 17.54).
- **Larger tumours:** T3 and T4 lesions are irradiated first. If the tumour persists after radiotherapy, excision of the entire lip may be necessary followed by PMMC flap reconstruction.
- Significant lymph nodes can be removed along with the primary tumour—**supraomohyoid block dissection**.

II. Radiotherapy

- It is indicated in all stages of carcinoma of the lip. Radiotherapy produces tumour necrosis resulting in a slow healing rate. Treatment lasts for several weeks and it delays the wound healing. Elderly patients who are not fit for surgery and carcinoma lip with fixed nodes are treated by irradiation.
- Commissure involvement is treated with RT than surgery.
- Dose: 4000–6000 centigray (cGy) units

Reconstruction of the lips

- There are various methods available to reconstruct the lip
- Up to 1/3rd of the lip can be sacrificed with direct closure
- Details are given in page 289 and summary (Table 17.2).

Please note: Students are advised to refer plastic surgery books for more details. Knowledge of some of these flaps will help you in getting more marks in the examinations.

Table 17.2 Reconstruction following resection of oral malignancy. Ca – carcinoma

Anatomic site of cancer	Surgery	Reconstruction
1. Ca lip (less than 2 cm)	V or W excision	Direct closure
2. Ca lip (large lesion)	Wide excision	**Bernard rotational flap**; Advancement flap
3. Advanced Ca lip	Total excision of lip (chin also)	Total lip reconstruction by using forearm flap
4. Small lesions lateral tongue	Partial glossectomy	Primary closure or secondary healing
5. Large defect tongue (> 2 cm)	Partial glossectomy	Reconstruction by radial forearm flap
6. Advanced lesions tongue	Total glossectomy	Reconstruction by using rectus abdominus flap
7. Floor of mouth T1 or T2	Wide excision	Radial artery forearm flap
8. Floor of mouth T3 or T4	Wide excision with removal of anterior mandible	Fibula flap or iliac crest graft—deep circumflex iliac artery (**DCIA**) is used
9. Buccal mucosa	Wide excision with removal of underlying buccinator muscle also	Direct closure or radial artery forearm flap
10. Lower alveolus	Wide excision with segmental resection of mandible	Primary reconstruction with fibula flap for edentulous mandible and DCIA for dentate mandible.

LIP RECONSTRUCTION (Figs 17.53 to 17.58)

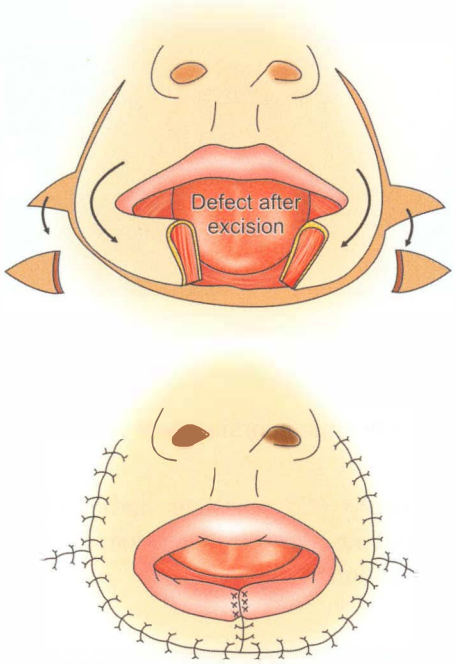


Fig. 17.53: Karapandzic flap—used for defect up to 2/3rd of lip size. Good functional result, good colour and texture match

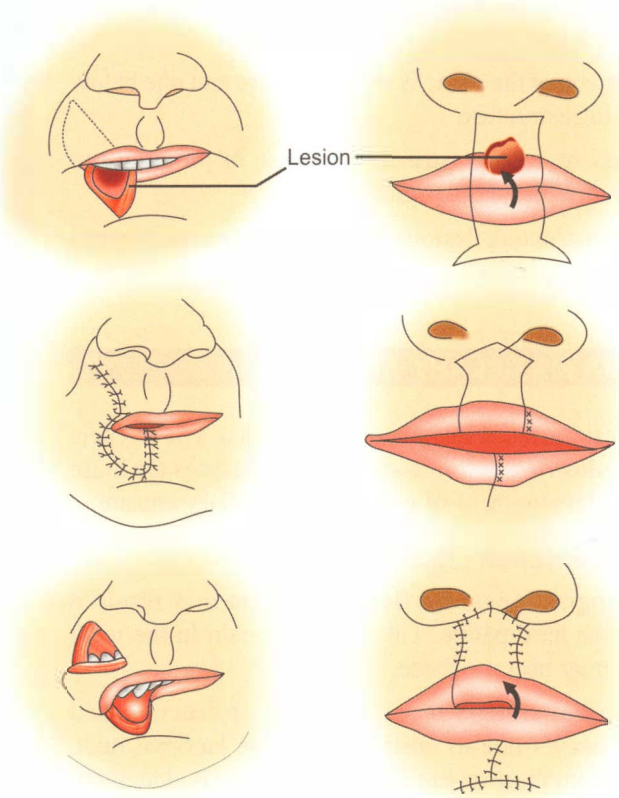
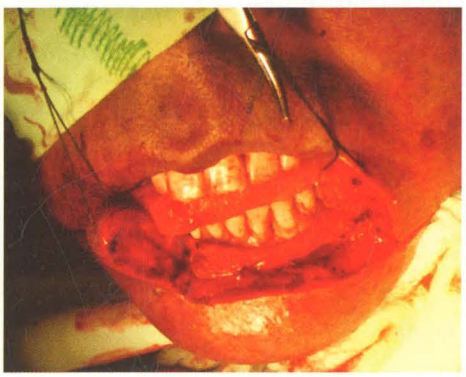
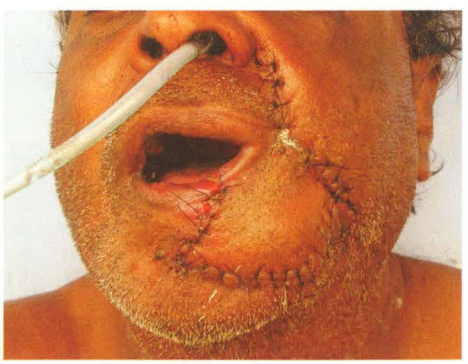


Fig. 17.54: Estlander flap. Used for lateral lip defects up to 1/3rd size. Good functional result

Fig. 17.55: Abbe's flap. Used for up to 1/3rd lip defect. Can be modified for both upper and lower lip. Produces good functional and cosmetic result





Figs 17.56 to 17.58: Wide excision of lip followed by reconstruction using nasolabial flap (*Courtesy:* Dr Biswas, Dr JK Jha, Dr SP Nayak, Chittaranjan National Cancer Institute (CNCI), Kolkata)

OHNGREN's line

- Antral carcinoma can be divided by a line joining medial canthus of eye to the angle of mandible. This line is called as Ohngren's line
- The region above the plane is suprastructure and one below is termed infrastructure
- Poor prognosis in suprastructure because of proximity to skull base and pterygoid region
- These suprastructure lesions as usually inaccessible and unresectable.

CARCINOMA MAXILLARY ANTRUM (Key Box 17.16)

It is rare in western countries but common in Asia. The workers in furniture industries, chromic and nickel industries are more prone for the development of carcinoma maxillary antrum.

Clinical presentation (Table 17.3)

1. Growth originating on the **floor of antrum** may result in bulge of the hard palate. This results in pain in the teeth and they may become loose.
2. When **medial wall** is involved, nasal obstruction and epiphora occurs due to obstruction of the lacrimal duct. Bleeding from the nose can also occur, if there is ulceration.
3. If **anterolateral wall** is involved, asymmetry of the face results in pain in the cheek. Anaesthesia over the skin of the cheek including upper lip occurs due to involvement of infraorbital nerve, a branch of maxillary division of the trigeminal nerve.
4. If the **roof** is invaded, proptosis and diplopia occurs.
5. **Posterior extension of the growth** is difficult to assess clinically. When it involves the pterygoid muscles, it results in trismus. Paraesthesia over the cheek, gums, lower lip, postnasal discharge are the other features of these tumours. They carry poor prognosis because of late presentation.

Fig. 17.59: Maxillary antral carcinoma

Lymphatic spread

- Nodal metastasis are uncommon in sinonasal malignancies.

Investigations

1. **Computed tomography** (CT scan) can define a lesion, its extent, bony destruction, posterior extension, etc. Hence, it is the first investigation of choice.

KEY BOX 17.16

CARCINOMA MAXILLARY ANTRUM

- Majority are squamous cell carcinoma
- Most important method of spread is contiguous spread
- Early presentation is rare because maxillary antrum serves no important function
- Tissue diagnosis is by biopsy of the mass protruding through nasal cavity/oral cavity or transnasal needle through medial wall of the maxilla.
- CT scan for bone involvement and MRI for soft tissue involvement are investigations of choice

Table 17.3 Clinical examination of a case of carcinoma maxillary antrum

• Palatine bulge	→ Growth in the floor
• Unilateral nasal obstruction (He is asked to breathe, closing the nostrils one by one)	→ Growth in the medial wall
• Asymmetry of the face	→ Growth expanding anteriorly, superficial surface
• Change in the level and sharpness of inferior orbital margin, proptosis	→ Growth on the roof (orbital surface)
• Swelling (indurated) temporal region	→ Posterior extension into infratemporal region and then into temporal region.
• Hard, fixed or mobile submandibular nodes	→ Metastasis

2. **Sinoscopy:** Fenestration will provide tissue for biopsy followed by curettage to reduce the tumour bulk and to drain necrotic contents outside.

Treatment

- **Radiotherapy is the main modality** of treatment in carcinoma maxillary antrum. Curative rate is around 70% in early cases. In advanced cases, radiotherapy is given first. This reduces the tumour bulk so that an unresectable lesion becomes resectable and maxillectomy can be done.
- Surgery can be done in the form of **total maxillectomy** when the growth involves entire maxilla or it is of **high grade** followed by postoperative radiotherapy.
- Tumours of the **lower half** of the antrum are treated by **partial maxillectomy**. It includes removal of the entire hard palate, alveolus and medial wall of the antrum up to and including middle turbinate.
- Indications and contraindications for surgery and radiotherapy are in similar lines as discussed earlier in this chapter.

NASOPHARYNX—CANCER

- Carcinoma arises in a small anatomic site bordered by nasal fossae, posterior wall continuous with posterior wall of oropharynx, body of the sphenoid and basilar part of occipital bone and soft palate.
- > 90% are squamous cell carcinoma out of which 40–50% are **undifferentiated (lymphoepithelioma)** and 5% are lymphomas.
- Incidence is higher in Asia–Southern China, Malaysia, etc.
- May be associated with **Epstein-Barr virus (EBV)**
- Present as **high posterior cervical lymphadenopathy**
- **Cranial nerve syndromes** are common due to tumour invasion of base of skull.
  1. **Retrosphenoidal syndrome** from involvement of cranial nerves II through VI manifests as unilateral ophthalmoplegia, trigeminal neuralgia, ptosis, etc.
  2. **Retroparotid syndrome:** Occurs due to compression of cranial nerve IX through XII and causes various symptoms depending on nerve involvement including Horner's syndrome.
- Diagnosis by endoscopy, biopsy, CT and MRI.
- Mainly managed by radiotherapy and chemotherapy

KEY BOX 17.17

PAINFUL ULCERS IN THE TONGUE

- Aphthous ulcers
- Dental ulcers
- Tubercular ulcers

KEY BOX 17.18

PAINLESS ULCERS IN THE TONGUE

- Carcinomatous ulcers
- Gummatous ulcers
- Systemic diseases

BENIGN LESIONS IN THE ORAL CAVITY

Differential diagnosis of ulcer in the tongue

(Fig. 17.60 and Key Boxes 17.17 and 17.18)

1. Aphthous ulcer

- Small, multiple, very painful ulcers, can occur at any age group. More common in females at the time of menstruation. These are called as **minor aphthous ulcers**.
- When they are larger, deeper and painful, they are called **major aphthous ulcers.**
- They are due to viral infection. These ulcers are superficial ulcers with erythematous margin.
- They subside within a few days. Temporary relief can be obtained by applying **salicylate gel.**
- Vitamin B complex is usually given for the satisfaction of the patients.

2. Dental ulcer

- These ulcers occur due to broken tooth, sharp tooth, ill-fitting dentures, prosthesis, etc. They are very painful ulcers.
- Such ulcers are common on the lateral margin and they heal when the tooth is removed. This is an example for traumatic ulcer. It should not be confused with carcinomatous ulcer which commonly occurs on the lateral margin.

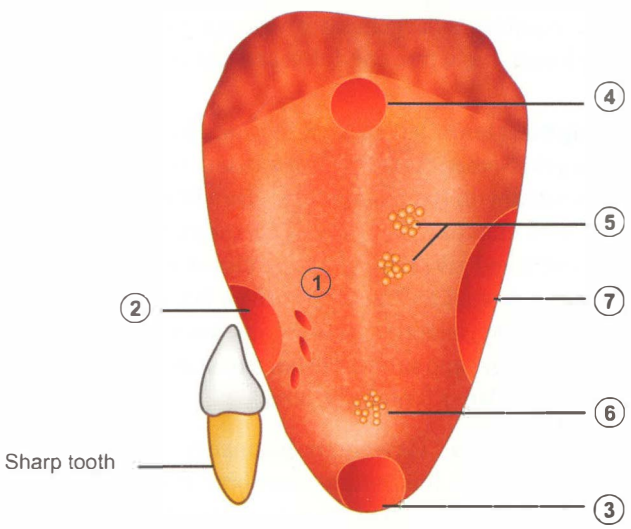


Fig. 17.60: Ulcers of the tongue—*see* text for numbers

3. Tubercular ulcer of tongue

- Tuberculosis affects tip of the tongue. These ulcers are very painful with enlargement of regional nodes.
- It occurs in patients with fulminating pulmonary tuberculosis.
- Ulcers have undermined edges. These ulcers are sometimes multiple with serous discharge.

4. Gummatous ulcer

- Gumma is a complication of tertiary syphilis resulting in a firm swelling in the midline in the anterior 2/3rd of the tongue. Induration is absent. Ulcer is nontender. Severe endarteritis obliterans results in the necrosis of gumma giving rise to gummatous ulcer. It has punched out edges and wash leather slough on the floor. Other sites of gumma include testis, palate, clavicle and liver. These ulcers are rare these days.

5. Systemic diseases

- Pemphigus
- Systemic lupus erythematosus (SLE)
- Lichen planus

6. Post-pertussis ulcer

- It occurs in children due to repeated coughing. Typical location of the ulcer on the under surface of the tongue and on the frenulum clinches the diagnosis.

7. Carcinomatous ulcer

- Lateral margin
- Nonhealing ulcer
- Everted edge
- Edge and base are indurated
- Bleeds on touch
- Fixity
- Significant lymph nodes in the neck.

MACROGLOSSIA

Diffuse painless enlargement of the tongue is described as macroglossia. It is a rare condition and can occur due to various causes.

1. **Lymphangioma:** In this condition, the tongue diffusely enlarges. Sometimes, it is a localised swelling. It may be associated with lymphangiomas elsewhere in the body such as cheek mucosa, lips, etc. The tongue becomes larger, indurated and gives rise to severe discomfort to the patients. Due to repeated trauma, the surface becomes ulcerated. It is treated by injecting sclerosants such as ethanolamine oleate, hypertonic saline. Partial excision may be necessary in cases of large lymphangioma.
2. **Haemangioma:** Cavernous haemangiomas occur in the tongue, lips, etc. It is present since birth but manifests during childhood. It presents with soft, cystic, fluctuant swelling and at times, pulsatile. Trauma due to teeth or food results in bleeding. Haemangioma of the tongue is treated on the same lines as lymphangioma. It is much more difficult to excise it, especially a large haemangioma. Preoperative angiography and ligation of lingual artery on both sides is necessary.
3. **Neurofibroma:** It may be associated with von Recklinghausen's disease. It is treated by hemiglossectomy.
4. **Muscular macroglossia:** This condition, though rare, is seen in cretins. The tongue is thickened and cannot be held in place. Hence, it protrudes outside. It is treated by partial excision.

SYPHILITIC LESIONS OF THE TONGUE

1. **Primary syphilis:** Primary chancre that occurs in the tongue is highly contagious. It affects the tip of the tongue. It produces a painful ulcer with large significant enlargement of regional lymph nodes.
2. **Secondary syphilis** produces a white patch in the tongue, lips and on the anterior pillars of fauces. In the tongue, these are multiple which coalesce to form **snail track ulcers**. The ulcers heal with **fine tissue paper scar**. In some cases syphilitic organisms produce a flat, hypertrophied epithelium which is described as **condyloma.** This is called as **Hutchinson's wart.**
3. **Tertiary syphilis:** It produces gumma. Syphilis also produces **chronic superficial glossitis**, which is characterised by bald tongue with loss of papilla and fissured tongue. **It is a precancerous condition.**

ODONTOMES

Definition: Odontomes are the cysts, malformations arising from epithelial or mesothelial elements of tooth resulting in swelling of the jaw. As a developmental anomaly, few epithelial cells proliferate, persisting as **epithelial debris of Mallasez**. Three important odontomes have been given in Table 17.4.

DENTAL CYST: RADICULAR CYST/PERIAPICAL CYST

Pathogenesis

This arises from a normally erupted, chronically infected, pulpless caries tooth. The caries tooth produces a low grade, chronic inflammation which stimulates epithelial debris to proliferate. Later this brings about degeneration of epithelial and mesothelial cells resulting in a cyst within the maxilla.

Clinical features

- Common in women around 3rd–4th decades
- Commonly affects the upper jaw (maxilla)

- It presents as a slow-growing swelling in the maxillary region resulting in deformity of the face.

Diagnosis

- Presence of caries tooth with expansion of maxilla
- X-ray—large, unilocular cyst in maxilla or orthopantomogram showing cyst in the mandible
- Aspiration of the cyst demonstrates cholesterol crystals.

Treatment

Excision of cyst with its epithelial lining through intraoral approach. After excision of the epithelium the cyst wall should be curetted, followed by soft tissue 'push-in' to obliterate dead space.

DENTIGEROUS CYST: FOLLICULAR ODONTOME

- Common in lower jaw (mandible) in women 30–40 years (Fig. 17.61).
- It occurs in relation to unerupted, permanent, molar tooth, most commonly the upper or lower third molar tooth.
- This unerupted tooth constantly irritates the cells, produces degeneration of the cells resulting in a dentigerous cyst. The cyst is lined by squamous epithelium surrounded by connective tissue. Within the cyst, the tooth lies obliquely or sometimes is embedded in the wall of the cyst. As it grows further, the cyst displaces the tooth to which it is attached. Thus, the tooth is displaced deeper and deeper and prevented from eruption.

Clinical features

- Absence of molar tooth

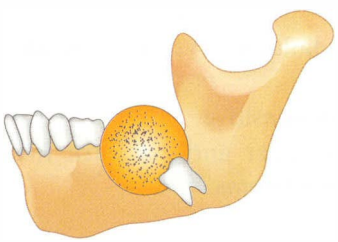


Fig. 17.61: Dentigerous cyst

Fig. 17.62: X-ray of the dentigerous cyst

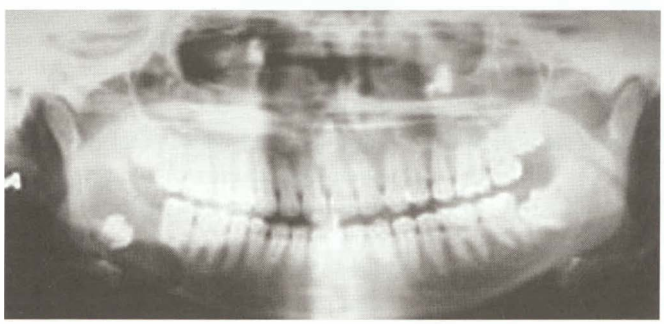


Fig. 17.63: Orthopantomogram showing unerupted tooth

- Expansion of mandible—since the inner table of the mandible is strong, the expansion mainly occurs in the outer aspect of mandible. The bone gets thinned out resulting in **egg-shell crackling**.

Diagnosis

X-ray mandible (Figs 17.62 and 17.63)

1. Tooth in the cyst
2. Soap-bubble appearance due to multiple trabeculations of the bone
3. Radiolucent well-defined swelling.

Treatment

- Small cyst—excision of the cyst by intraoral approach.
- Large cysts—managed by marsupialisation.

ADAMANTINOMA

- It is also called multilocular cystic disease, ameloblastoma, Eve's disease (Fig. 17.64).
- This tumour arises from ameloblasts (enamel forming cells).
- It is a benign tumour, very slow-growing and behaves like a basal cell carcinoma. Inadequate treatment results in local recurrence and later metastasis, Hence, even though the tumour is benign, it has to be treated like malignant tumour.

Sites

- **Mandible** is the most common site
- **Tibia** is the 2nd common site. It can be explained by inclusion of abnormal embryonic epithelium.
- **Pituitary** is another common site where adamantinoma can occur. **Both pituitary stalk and enamel arise from oral epithelium.**

Clinical features

- Patients in 4th or 5th decade are commonly affected.
- This is a slow growing jaw tumour in the region of angle of mandible and horizontal ramus of the mandible.

Fig. 17.64: Adamantinoma left jaw

- As the tumour grows, it undergoes cystic degeneration resulting in multiple cystic spaces. Hence, it is called multilocular cystic disease.
- As it grows it causes expansion of the outer table of the mandible and causes fracture mandible.
- Patient may present with complaints of falling teeth.

Diagnosis

X-ray: A large cyst and small multiple cysts due to the trabeculations giving it a 'honeycomb' appearance.

Treatment

- Even though it is **benign, simple curettage or enucleation may result in recurrence** and chances of recurrent adamantinoma turning into malignancy are high. Hence, wide excision with 1 cm of healthy normal tissue should be removed. It may amount to **segmental excision of the mandible or hemimandibulectomy** (Key Box 17.19 and Table 17.4).

KEY BOX 17.19

ADAMANTINOMA

- Locally invasive solid tumour
- Nonfunctional, **intermittent in growth,** unicentric, **multilocular**
- Spreads within the medullary bone
- Invades soft tissues
- Should not fragment the tumour cells
- **Subperiosteal excision** should **not be done** as it may result in recurrence
- **Incomplete excision results in recurrence** and metastasis to the lung
- Hence, even though it is a **benign tumour** it is treated by wide **excision** or hemimandibulectomy.

Differential diagnosis

1. Giant celled reparative granuloma (Jaffe tumour

It is a benign tumour which occurs due to haemorrhage within the bone marrow.

Pathology

- It affects antral part of maxilla or mandible causing enlargement of the jaw.
- Stroma is vascular consisting of thin-walled blood vessels scanty collagen, connective tissue cells.
- Microscopic features mimic giant cell epulis or brown tumour of hyperparathyroidism.

Clinical features

- Unlike an adamantinoma, this tumour affects females in the age group of 10–25 years.
- Painless enlargement of the jaw is the presenting feature X-ray demonstrates radiolucent artery.

Treatment

- Calcitonin 0.5 mg (100 units) daily subcutaneous injection over a period of one year has been recommended as a first line of treatment. It has shown resolution of the tumour.
- Curettage is the surgical line of treatment.

2. Osteoclastoma

- This is a rare tumour seen in the lower jaw.
- Males between the age of 25 and 40 years are commonly affected.
- Unlike adamantinoma, it is a rapidly growing tumour.
- As the tumour enlarges, both tables of the lower jaw are thinned out.
- X-ray may show a large, radiolucent cyst with pseudo-trabeculation.
- Even though benign, it is radiosensitive.
- However, recurrence can occur and can turn into malignancy like that of adamantinoma.

Table 17.4 Comparison of three common odontomes

	Dental cyst	Dentigerous cyst	Adamantinoma
1. Aetiology	Caries tooth	Unerupted tooth	True neoplasm—ameloblasts
2. Site	Maxilla	Mandible	Mandible
3. Age	30–50 years	20–30 years	40–50 years
4. Palpation	Smooth, thin bone-eggshell crackling	Smooth, expansion of the outer table of mandible	Expansion of mandible in the 3rd molar region, cystic areas
5. X-ray	Radiolucent unilocular cyst	Radiolucent unilocular cyst with unerupted teeth	Large radiolucent area with fine honey-combing
6. Treatment	Subperiosteal excision	Subperiosteal excision	Wide excision even though benign
7. Spread	Does not occur	Does not occur	Local spread, recurrence, late spread to the lung are the features

EPULIS

- *Epulis* means "upon the gum". It refers to solid swelling situated on the gum (Key Box 17.20).
- It arises from alveolar margin of the jaw.
- Very often patients present with swelling on the gum which is painless.

Types

Granulomatous epulis (Fig. 17.65)

- Precipitating factors are caries tooth, dentures, poor oral hygiene.
- It manifests as a mass of granulation tissue around the teeth on the gums. It is a soft to firm, fleshy mass and bleeds on touch.
- Pregnancy epulis refers to this variety (gingivitis gravidarum).

Fibrous epulis (Fig. 17.66)

- It is the commonest form. A simple fibroma arising from periodontal membrane, presents on the gum. It may undergo sarcomatous change. It is a firm polypoidal mass, slowly growing and nontender.

Giant cell epulis

- It is also called myeloid epulis.
- It is an osteoclastoma arising in the jaw. It presents as hyperaemic vascular, oedematous, soft to firm gums with indurated underlying mass due to expansion of the bone. It may ulcerate and result in haemorrhage. X-ray shows bone destruction with ridging of walls (pseudo-trabeculation).
- Small tumours are treated by curettage.
- Large tumours are treated by radical excision.

KEY BOX 17.20

EPULIS

• Soft epulis	– Granulomatous
• Firm epulis	– Fibrous
	– Giant cell
• Hard epulis	– Carcinomatous
• Malignant epulis	– Carcinomatous
• Dangerous epulis	– Fibrosarcomatous

Carcinomatous epulis

- This is an epithelioma arising from mucous membrane of the alveolar margin.
- Typically, it presents as a nonhealing, painless ulcer. It slowly infiltrates the bone.
- Hard regional lymph nodes are due to metastasis.
- Treated by wide excision which includes removal of segment of the bone.

MEDIAN MENTAL SINUS

This is a sinus in the midline just beneath the mentum.

Aetiopathogenesis

It is produced by an apical abscess of lower incisors which penetrate buccal cortical plate below the origin of mentalis muscle. This muscle takes origin from labial surface of alveolar process just above the labial sulcus. Hence, pus discharges through a sinus in the centre of chin.

Clinical presentation (Fig. 17.67)

- Patients present with recurrent swelling in the submental region which bursts open spontaneously discharging at times mucus and seropurulent fluid.
- Repeated history of swelling, discharge and healing are common presentations.

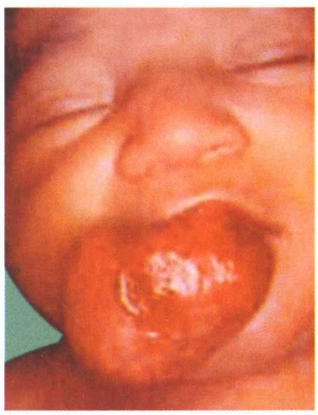


Fig. 17.65: Granulomatous epulis

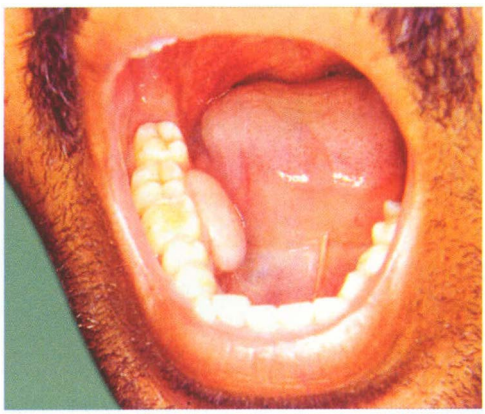


Fig. 17.66: Fibrous epulis

Fig. 17.67: Median mental sinus

CLINICAL NOTES

We had a patient with discharging sinus in the region of mentum for 1½ years who had seen many practitioners including a surgeon who curetted the lesion twice. However, the lesion reappeared soon. This case is a classical example of what the mind does not know, eyes cannot see.

- Diagnosis is established by examination of the oral cavity. which reveals evidence of caries tooth.

Treatment

Once the caries tooth is extracted, sinus will heal spontaneously.

VINCENT'S ANGINA

It is an acute ulceromembranous stomatitis or acute ulcerative gingivitis and stomatitis. The disease is caused by Vincent's organisms—*Borrelia vincentii*, an anaerobic spirochaete and fusiformis. These are gram-negative rods which are the normal pathogens of oral cavity.

Precipitating factors

- Malnutrition, diabetes mellitus, caries tooth, warm seasons, winter, etc.
- The disease starts in the intergingival defects as a deep penetrating ulcer which results in a **spontaneous gingival haemorrhage**. There is a thick membrane covering the ulcer.
- Once infection spreads to tonsillar region, it is called as Vincent's angina—very severe painful condition.

Clinical features

- Common in children and young adults between 20 and 40 years of age.
- It presents with very painful gums with fever, malaise and toxaemia.
- Gums are swollen, red, inflamed with or without slough.
- Difficulty in swallowing, painful swallowing (odynophagia), **foetor oris**, features of toxaemia and high grade fever are characteristic of this condition.

Treatment

- Improve nutrition. Mouth washes with hydrogen peroxide help in washing away the membrane. Injection Penicillin: 10 lakh units, IM 6th hourly for 6–7 days.
- Since they are anaerobic organisms, metronidazole 400 mg thrice/day for 7–10 days, should be given.

CLEFT LIP AND CLEFT PALATE

CLEFT LIP

- Cleft lip results from abnormal development of the median nasal and maxillary process.
- Cleft palate results from a failure of fusion of the two palatine processes.

Types of cleft lip (Figs 17.68 and 17.69)

I. **Central:** It is very rare and occurs due to failure of fusion of two median nasal processes.

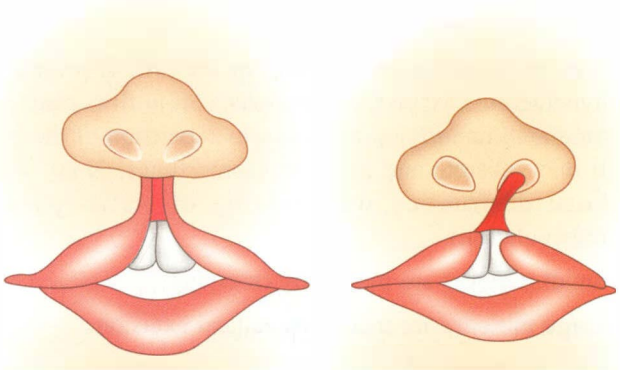
II. **Lateral:** It is the commonest variety wherein there is a cleft between the frenulum and the lateral part of the upper lip. This is due to imperfect fusion of maxillary process with median nasal process. The lateral variety can be unilateral or bilateral.

III. **Complete or incomplete:** In cases of complete variety, cleft lip extends to the floor of the nose. In cases of incomplete variety, the cleft does not extend up to the nostril.

IV. **Simple or compound:** Compound refers to cleft lip associated with a cleft in the alveolus.

Clinical features (Figs 17.70 and 17.71)

1. In 80% of the cases, cleft lip is unilateral and in about 60% of the cases it is associated with cleft palate.



Figs 17.68 and 17.69: Types of cleft lip (I and II)

Figs 17.70 and 17.71: Cleft lip (*Courtesy:* Dr CG Narasimhan, Senior Consultant Surgeon, Mysore, Karnataka)

2. In many cases, nostril is widened.
3. Maldevelopment or malalignment of the teeth in relation to the cleft is common.

Functional effect

1. Presence of cleft lip does not interfere much with sucking. However, there may be some difficulty in bottle feeding.
2. Some degree of difficulty in speech (disarticulation) is present.

CLEFT PALATE

Development of palate

- Palate is developed around 6–8 weeks of intrauterine life from **3 components**. The premaxilla is developed from the median nasal process and maxillary process contributes one palatine process on each side.
- The line of fusion of these processes is in the form of a letter Y.
- Imperfect fusion or developmental anomalies results in cleft palate.

Types (Fig. 17.72)

I. **Complete:** Failure of fusion of palatine processes and premaxilla results in complete cleft palate. In such situations, the nasal cavity and mouth are interconnected. When premaxilla is not fused with both palatine processes, it hangs down from the septum of nose. Thus, complete cleft can be of two types as shown below in the diagram.

II. **Incomplete:** When the fusion of three components of palate takes place, it starts from uvula and then backwards. Thus, various types of incomplete fusion results.
   - Bifid uvula.
   - The whole length of soft palate is bifid.
   - The whole length of soft palate and the posterior part of hard palate are involved. On the other hand, anterior part of palate is normally developed. In about 25% of cases, cleft palate alone and in 50% of cases, both cleft palate and cleft lip are encountered.

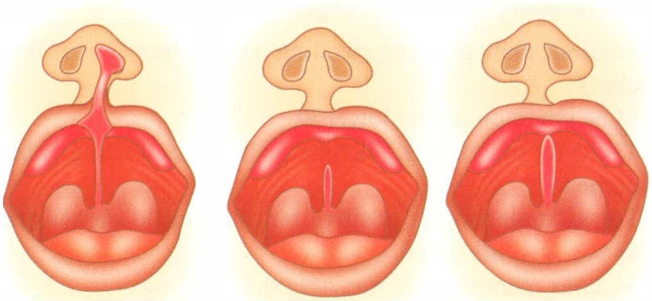


Fig. 17.72: Types of cleft palate (I, IIa and IIb)

Effects of cleft palate

1. Presence of cleft palate interferes with swallowing to some extent.
2. They are unable to pronounce the consonant sounds such as B, D, K, P, T.
3. **Teeth:** Upper lateral incisors may be small or even absent. The maxilla tends to be smaller. Teeth are crowded.
4. **Nose:** Oral organisms contaminate the upper respiratory mucous membrane through cleft palate.
5. **Hearing:** Even with repair, acute and chronic otitis media and hearing problems can occur.

Management of cleft lip and palate

- **A multidisciplinary approach** involving plastic surgery, orthodontics, speech pathology, ENT department, prosthodontics and paediatrics department is needed to rehabilitate the cleft palate cases. This approach to the problem results in aesthetically acceptable end result without much functional deficiencies.
- **Feeding advice:** Cleft palate babies are unable to suck mothers' milk because intraoral negative pressure cannot be created due to communication between oral and nasal cavity. Thus, expressed mother's milk is given by spoon with head end of baby elevated by 45 degrees. Swallowed air during feeding is released frequently by burping.

Cleft lip repair (Key Box 17.21)

Timing: Majority of surgeons follow "RULE OF 10" as a guide for timing of lip and anterior palate repair. At the time of repair, haemoglobin should be more than 10 g%, age approximately 10 weeks, weight more than 10 lb (4.54 kg) and total leukocyte count less than 10,000/cu mm (i.e. no infection).

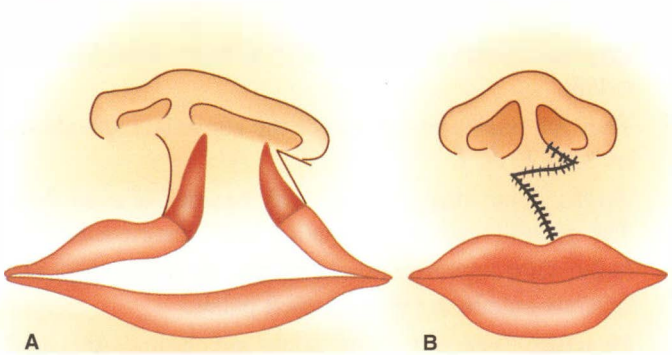
KEY BOX 17.21

RULE OF 10

- Hb > 10 g%
- Age approx : 10 weeks
- Weight > 10 lb (4.54 kg)
- TC < 10,000/mm^3

Types of cleft lip repair

- For unilateral cleft lip repair most commonly used methods are Millard rotation advancement flap and Tennison-Randall triangular flap method.
- Bilateral cleft lip can be repaired in single stage or in two stages at the interval of 3–6 months. For two stage repair, any one of the methods described for unilateral cleft lip can be used. For bilateral repair in one stage, Veau III method is simple and gives satisfactory results. Other single stage methods which give good results are **Millard's single stage** procedure and Black procedure.



Figs 17.73A and B: Basic steps of lip repair

Basic steps of lip repair

- Markings are made according to the method selected (e.g. Millard's repair, Fig. 17.73A and Tennison-Randall repair, Fig. 17.73B).
- Adrenaline-saline solution (1:200,000) is injected in the lip and labial sulcus for haemostasis.
- Full thickness of lip is incised along the marking.
- Lip repair is done in three layers—mucosa, muscle and skin. For better aesthetic result, Cupid's bow should become horizontal, **white line** continuity should be repaired and there should be no vermilion notching.

Cleft palate repair

Timing: Early repair results in retarded maxillary growth due to surgical trauma to growth centre and periosteum. Delay in repair results in speech defect. Best balanced result is achieved by repairing between one and a half years.

Types

- Palate repair: Palate is repaired by palatal shelves. **Mucoperiosteal flaps raised from various methods are available for palate repair**.
- Most commonly used method is V-Y, pushback palatoplasty.

Steps of 'V-Y' pushback palatoplasty (Fig. 17.74)

- Palate is infiltrated with 1 : 2 lakh adrenaline-saline solution.
- Two mucoperiosteal flaps are elevated, one from either side of palatal shelves. Then, nasal layers are mobilised.
- Palate is closed in three layers—nasal layer, muscle layer, oral layer.
- In V-Y push back palatoplasty, palatal lengthening is achieved by V-Y plasty. Hook of hamulus can be fractured to relieve tension on suture line by relaxing the tensor palati muscle.

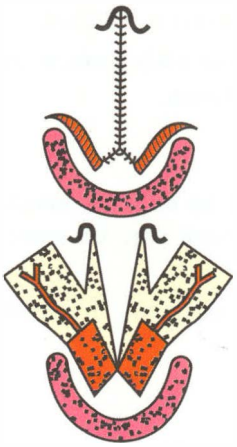


Fig. 17.74: V-Y palatoplasty

MISCELLANEOUS

ECTOPIC SALIVARY GLAND TUMOUR

- **Palate** is the most **frequent site**
- These tumours can also occur anywhere in the mouth or pharynx
- **It is a slow-growing and painless tumour.** At this stage, it may feel firm or hard without ulceration of mucous membrane.
- Slowly it **ulcerates.** Thus, it can have the shape of a **'verrucous'** carcinoma. Such lesions are ulcerated, hard, and painful with irregular margins.
- They are of **low-grade malignancy.**
- Neglected cases can invade base of skull and spread to lymph nodes.
- **Wide excision** with or **without reconstruction** is the treatment of choice.

MUCOUS CYSTS

- They are examples of retention cyst
- They occur due to obstruction of the duct of many mucous secreting glands which cover the inner surface of the lips and whole of inside of mouth (Fig. 17.75).
- They are also a type of **extravasation** cyst.

Clinical symptoms and signs

- Painless, slow growing swelling on the inner side of lip or cheek.
- Most common on the lower lip
- Typically round, soft, fluctuant, pale pink swelling or blue domed.
- Transillumination is positive but difficult to demonstrate as the cysts are small.
- Mucous membrane is free over the swelling.

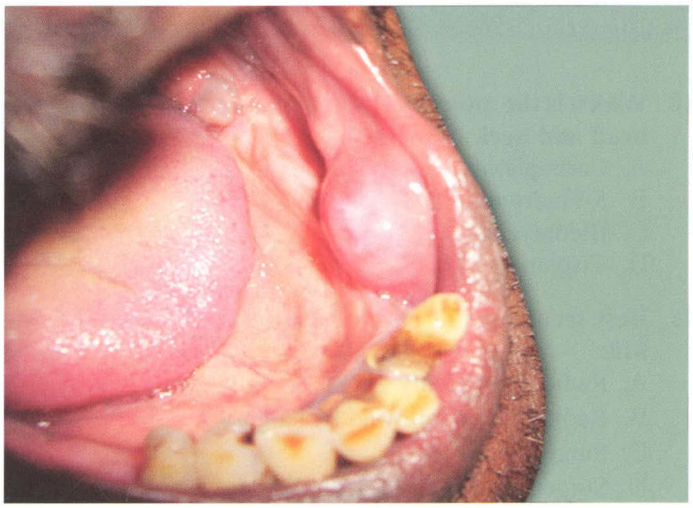


Fig. 17.75: Mucous cyst in the floor of the mouth

Differential diagnosis

- **Pyogenic granuloma:** It is red in colour, soft and bleeds. It may be associated with trauma or persistent infection.
- **Ectopic salivary gland tumours:** They are firm and non-tender swellings.

Treatment

- Excision can be done under local anaesthesia
- Once mucous membrane is incised, swelling can be dissected all around separating it from orbicularis oris/ buccinator muscle and it is removed.
- The mucous membrane is closed with absorbable sutures.

A case of malignant melanoma of the oral cavity

(*see* the clinical notes below)

CLINICAL NOTES

This 48-year-old man (Figs 17.76 to 17.78) *presented with an innocent looking, painless submandibular lymph node enlargement. It was firm and nontender. Initially, it was thought to be due to dental caries. Examination of the floor of the mouth revealed an interesting melanomatous lesion. Diagnosis was malignant melanoma with metastasis in nodes. This is just to remind that melanoma can also occur in the oral cavity (mucosa and firm skin).*

Fig. 17.76: This patient presented with right submandibular lymph node enlargement

Fig. 17.77: Oral cavity examination reveals a pigmented ulcerated lesion

Fig. 17.78: Closeup view reveals it is a malignant melanoma

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All the topics have been updated
- New photographs and key boxes have been added
- Reconstruction of the oral cavity has been dealt in detail including lip
- PMMC flap has been discussed in more detail.

MULTIPLE CHOICE QUESTIONS

1. Which one of the following has highest chance of malignancy?
A. Acanthosis B. Dyskeratosis
C. Leukoplakia D. Speckled leukoplakia

2. Drug used to treat leukoplakia is:
A. Vitamin E B. Isonicotonic acid
C. Isotretinoin D. Niacin

3. Following are true for verrucous carcinoma *except:*
A. Very slow growing
B. Poorly differentiated carcinoma
C. Spread by lymphatics is not common
D. Surgery is the best treatment

4. Carcinoma tongue spreads to following lymph nodes *except:*
A. Submandibular
B. Submental
C. Jugulo-omohyoid
D. Supraclavicular node

5. Following malignant lesions in the posterior third of the tongue can occur *except:*
A. Squamous cell carcinoma
B. Adenocarcinoma
C. Lymphoepithelioma
D. Sebaceous carcinoma

6. The investigation of choice in oral cancer to find out the skull base involvement is:
A. CT scan
B. Angiography
C. MRI
D. Ultrasound

7. Which cancer in the oral cavity — T1 N0 M0 – requires supraomohyoid block dissection?
A. Carcinoma buccal mucosa
B. Verrucous carcinoma lower lip
C. Carcinoma upper lip
D. Carcinoma floor of the mouth

8. Following are true for chronic hyperplastic candidiasis *except:*
A. Invasion of *Candida albicans*
B. Antifungal treatment helps
C. High malignant potential
D. Floor of the mouth is affected

9. Which one of the following condition has high incidence of distant spread?
A. Carcinoma buccal mucosa
B. Carcinoma tongue
C. Carcinoma floor of the mouth
D. Nasopharyngeal carcinoma

10. Which is the most effective chemotherapeutic drug for head and neck cancers?
A. Carboplatins
B. 5-Fluorouracil
C. Bleomycin
D. Cisplatin

11. Best treatment of carcinoma buccal mucosa–T1 N0 M0:
A. Radiotherapy only
B. Radiotherapy followed by surgery
C. Surgery only
D. Surgery followed by radiotherapy

12. Khaini chewers more susceptible for development of which carcinoma?
A. Carcinoma lip
B. Cracinoma tongue
C. Carcinoma buccal mucosa
D. Carcinoma floor of the mouth

13. Estlander flap is used to cover:
A. Central defects of lip—lower lip
B. Central defects of lip—upper lip
C. Lateral defects of more than 50%
D. Lateral defects up to 30%

14. Following are true for precancerous lesions of lip/oral cavity *except:*
A. Keratoacanthoma
B. Leukoplakia
C. Erythroplakia
D. Submucous fibrosis

15. Following are features of carcinoma maxillary antrum *except:*
A. Can cause asymmetry of face
B. Can cause proptosis
C. Can cause infraorbital nerve paralysis
D. Can cause buccal branch of fascial nerve paralysis

16. Following are the boundaries of nasopharyngeal space *except:*
A. Nasal fossae
B. Basilar part of occipital bone
C. Body of sphenoid
D. Cribriform plate of ethymoid bone

17. Following are true for nasopharyngeal carcinoma *except:*
A. Presents as high anterior cervical lymphadenopathy
B. Can present as trigeminal neuralgia
C. Compression on IX and X cranial nerves
D. Can present as ophthalmoplegia

18. Following are painless ulcers in the tongue *except:*
A. Gummatous ulcers
B. Carcinomatous ulcers
C. Systemic diseases
D. Tuberculous ulcers

19. Following are painful ulcers in the tongue *except:*
A. Gummatous ulcers
B. Tuberculous ulcers
C. Aphthous ulcers
D. Dental ulcers

20. Following are true for syphilitic lesions of the tongue *except:*
A. Snail track ulcers
B. Gumma
C. Hutchinson's wart
D. Hunterian chancre

21. Following are true for dental cyst *except:*
A. Upper jaw is commonly involved
B. It is a large unilocular cyst
C. Cyst contains cholesterol crystals
D. Arises from unerupted tooth

22. Which one of the following swelling does not contain cholesterol crystals?
A. Branchial cyst
B. Sebaceous cyst
C. Dental cyst
D. Hydrocoele

23. Following are true for dentigerous cyst *except:*
A. Upper jaw is commonly involved
B. Produces egg shell crackling
C. X-ray shows soap bubble appearance
D. Arises from unerupted tooth

24. Adamantinoma of the jaw has following features *except:*
A. It is a malignant tumour
B. It spreads within medullary bone
C. It is treated by wide excision
D. Mandible is the most common site

25. Which of the following is unilocular cyst?
A. Adamantinoma
B. Dentigerous cysts
C. Epididymal cyst
D. Dental cyst

ANSWERS

1 D	2 C	3 B	4 D	5 D	6 C	7 D	8 D	9 D	10 D
11 C	12 A	13 D	14 A	15 D	16 D	17 A	18 D	19 A	20 D
21D	22 B	23 A	24 A	25 D					

18

Salivary Glands

- Surgical anatomy
- Acute parotitis
- Submandibular sialoadenitis
- Salivary gland tumours
- Summary of malignant tumours
- Frey's syndrome
- Sjögren's syndrome
- Mikulicz disease
- Parotid fistula
- Surgery for facial nerve palsy
- Peripheral nerve repair and transfers
- What is new?/Recent advances

Introduction

There are three pairs of salivary glands—parotid, submandibular and sublingual. In addition to these, there are many (450) minor salivary glands located in the cheek, mucosa, lips, palate and base of the tongue. Parotid, the "big brother of 3", suffers mainly from three diseases—infection, enlargement and tumour. Submandibular salivary gland suffers from mainly two diseases—sialoadenitis and tumours. Other salivary glands are of minor importance. However, it should be remembered that the commonest tumour of minor salivary glands is malignancy.

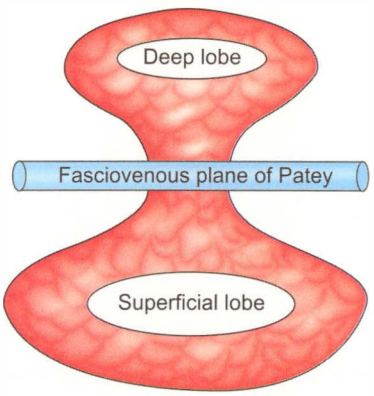


Fig. 18.1: Two lobes of parotid gland

SURGICAL ANATOMY OF THE PAROTID GLAND

Parotid gland is present on the lateral aspect of the face, divided by the facial nerve into superficial lobe and deep lobe. Superficial lobe overlies the masseter and the mandible. Deep lobe is wedged between the mastoid process and the styloid process, ramus of the mandible and medial pterygoid muscle.

The superficial lobe also receives a duct from the accessory lobe which is in the region of zygomatic arch/zygomatic process. The **duct of parotid, Stensen's duct**, 2–3 mm in diameter, receives tributaries from superficial, deep and accessory lobes, passes through the buccinator muscle and opens in the mucosa of the cheek **opposite the upper 2nd molar tooth.** Parotid gland is covered by a true capsule which is a condensation of fibrous stroma of the gland and a **false capsule** formed by **parotid fascia**, a part of the deep cervical fascia.

Facial nerve

After emerging from stylomastoid foramen, it hooks around the condyle of mandible, enters the substance of the parotid and divides into 2 major branches, **zygomaticotemporal** and **cervicofacial**. Facial nerve along with retromandibular vein (which is formed by the union of superficial temporal vein and maxillary vein, formed from branches of pterygoid plexus of veins) are present in this plane. This plane is called the **fasciovenous plane of Patey** (Figs 18.1 and 18.2). The facial nerve then gives rise to 5 branches which are interconnected

Table 18.1 Facial nerve and its branches

Branches of facial nerve		Muscles supplied
1. Temporal	→	Auricularis anterior and superior portion of frontalis
2. Zygomatic upper	→	Frontalis and upper half of orbicularis oculi
lower	→	Lower half of orbicularis oculi and muscles below the orbit
3. Buccal	→	Buccinator, orbicularis oris and a few fibres of elevators of the lower lip
4. Mandibular	→	Muscles of the lower lip
5. Cervical	→	Platysma

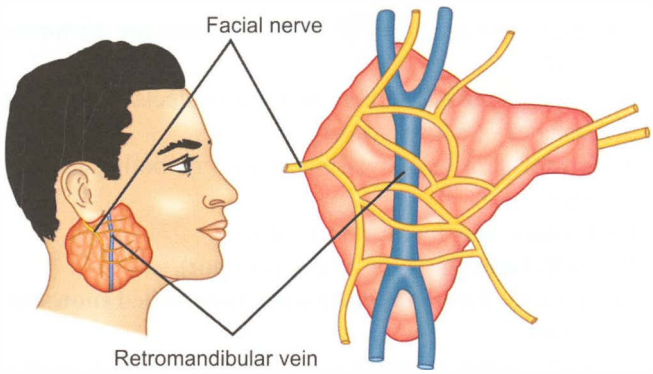


Fig. 18.2: Surgical anatomy of the parotid gland

like the foot of a goose, called ***Pes anserinus***. Branches of facial nerve in the face and the muscles supplied by these nerves are given in Table 18.1.

ACUTE PAROTITIS

Acute inflammation of the parotid can occur due to bacterial or nonbacterial causes. It can be unilateral or bilateral (Key Box 18.1). Three important causes and their treatment are given below:

1. **Mumps parotitis:** Mumps[1] is an acute generalised viral disease with painful enlargement of salivary glands, chiefly the parotids. The virus belongs to Paramyxoviridae family and only one serotype is known. The disease spreads from a human reservoir by direct contact, airborne droplets or fomites contaminated by saliva and possibly by urine (Fig. 18.3).

Clinical manifestation

- Incubation period is 10–24 days. Fever, headache and muscular pain are usually found. Both parotids are enlarged with pain and temperature.
- Swelling starts subsiding by 3–7 days of time.

[1]It causes parotitis, orchitis and pancreatitis.

KEY BOX 18.1

CAUSES OF ACUTE PAROTITIS

1. Viral	:	**Mumps**—Commonest Coxsackie A and B Parainfluenzae 1 and 3 Echovirus Lymphocytic choriomeningitis
2. Bacterial	:	Usually ascending infection *Staphylococcus aureus*
3. Recurrent parotitis of childhood	:	Recurrent, mistaken for mumps Resolves at puberty
4. Specific infections	:	Mycobacterial, cat-scratch disease, syphilis, toxoplasmosis
5. Allergic	:	Food and drugs
6. Sexual diseases	:	HIV-related
7. Postradiation	:	Reduction in the salivary juice
8. Postoperative	:	Due to dehydration.

Fig. 18.3: Acute parotitis due to viral infection

Treatment

- **If symptomatic:** Maintenance of good oral hygiene and hydration is useful. Antibiotics may be given to prevent secondary infection. One episode of infection confers lifelong immunity.

2. **Acute bacterial parotitis:** *Staphylococcus aureus* infection of parotid produces serious illness with marked engorgement of parotid. Typically, it produces parotid abscess. Diabetes, malignancy, malnutrition increase the risk. Decreased salivary secretion is an important predisposing factor.
3. **Reduction in salivary juice:** It can occur due to various factors mentioned in the box. **Postoperative parotitis** can be prevented by good mouth care and good oral hygiene. Due to poor oral hygiene, ascending infection occurs from the oral cavity resulting in parotitis (Key Box 18.2).

KEY BOX 18.2

CAUSES OF ↓ SALIVARY FLOW

- Postoperative
- Poor oral hygiene
- Dehydration
- Enteric fever, septicaemia
- Postradiotherapy, for oral cancer

Clinical features

- A patient who is recovering in the postoperative period may complain of pain and swelling in the parotid region. Presence of severe pain with a very sick, toxic look and high grade fever, chills and rigors indicates parotid abscess. **Diffuse brawny swelling is characteristic**.
- The swelling is due to inflammation of parotid and since it is enclosed by parotid fascia, the swelling takes the shape of parotid gland. However, it is not common for a parotid abscess to raise the ear lobule. For the reason mentioned above, fluctuation is a late feature. **If the abscess is not drained, it is likely to rupture into the external auditory canal** (Key Box 18.3).
- The opening of the parotid duct may be inflamed and on gentle compression of the parotid gland, pus can be seen coming out of the parotid duct.

KEY BOX 18.3

SWELLINGS WHEREIN ONE SHOULD NOT WAIT FOR FLUCTUATION

- Parotid abscess
- Breast abscess
- Ischiorectal abscess
- Pulp space infection
- Any deep seated abscess

Treatment

I. Conservative line of management

- Indicated in a stage of cellulitis with no abscess.
- Maintaining good hydration of the patient in the post-operative period.
- Improvement in the oral hygiene—mouth washes with potassium permanganate ($KMnO_4$) solution.
- Appropriate antibiotics against staphylococci, such as cloxacillin, is administered in the dose of 500 mg, 6th hourly along with metronidazole 400 mg, 8th hourly to treat anaerobic infections.
- It takes about 3–5 days for the inflammation to settle down.

II. Surgical treatment when there is pus

- Under general anaesthesia, an adequate vertical incision is made in front of the tragus of the ear up to deep fascia.

KEY BOX 18.4

DRAINAGE OF PAROTID ABSCESS

- Should not wait for fluctuation
- High grade fever, toxicity are indications
- Vertical incision
- **Hilton's method** is preferred to break multiple loculi

Open the deep fascia in two or three places and drain with blunt haemostat so as to avoid damage to facial nerve. This is described as **Blair's method** of drainage of parotid abscess. A drainage tube has to be kept which can be removed after 3–4 days (Key Box 18.4).

RECURRENT PAROTITIS OF CHILDHOOD

- Children between ages of 3 and 6 years are commonly affected.
- Aetiology is unknown, may be due to sialectasis (dilatation of branches of salivary duct)
- Recurrent pain and swelling of one or both parotids is common.
- Each attack may last for 3 to 7 days.
- It is **self-limiting** (if the attack is minor).
- Sialography shows **punctate sialectasis**, called **snowstorm** appearance.
- A short course of antibiotics has to be given to cover *Streptococcus viridans*.
- Rarely, superficial parotidectomy may be necessary (Fig. 18.4).

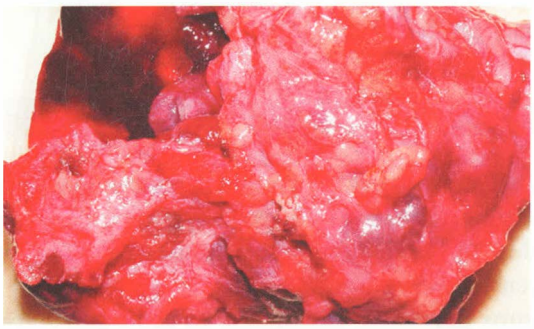


Fig. 18.4: Superficial parotidectomy for recurrent parotitis. Please remember **chronic parotitis in children is pathognomonic of HIV infection**

SURGICAL ANATOMY OF THE SUBMANDIBULAR SALIVARY GLAND (Fig. 18.5)

- Submandibular salivary gland is located in the sub-mandibular triangle. It lies partly below and partly above the mandible.
- It is in very close contact with the belly of the digastric muscle. At surgery, once the deep fascia is **opened, the intermediate tendon of digastric is located** and when it is retracted downwards, mobilisation of the gland becomes easy.

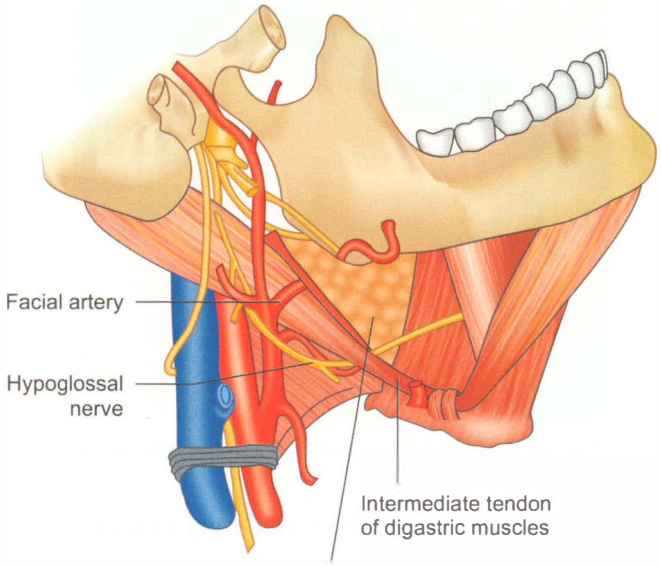


Fig. 18.5: Anatomy of the submandibular salivary gland

- Submandibular salivary gland is divided into a superficial and a deep part by the mylohyoid muscle which forms the oral diaphragm. During excision of the gland, a few fibres of mylohyoid are also removed. When submandibular salivary gland enlarges, **it is bidigitally palpable** because the **deep portion is deep to mylohyoid** and it is in the floor of the mouth.
- Facial artery enters the gland from its posterolateral surface and **deeply grooves the gland**. It is ligated at this place first during excision of the gland. After grooving the gland, it ascends laterally and curls around the lower border of mandible to enter the face. It is ligated at this place also.
- Main duct of submandibular gland, **Wharton's duct** arises from deep part of gland and opens on a papilla beside the frenulum of the tongue in the oral cavity.
- In a deeper plane, the gland is related to **two nerves—lingual and hypoglossal.**

CHRONIC SUBMANDIBULAR SIALOADENITIS

- Obstruction is the most important cause of submandibular sialoadenitis. Trauma to the floor of the mouth is another cause.
- Obstruction can be due to stone, disease, stricture of the duct, or fibrosis of the papilla (calculus – most common).
- The causative organism is Staphylococcus.

SIALOADENITIS DUE TO CALCULI

- The disease starts with acute bacterial sialoadenitis which occurs secondary to obstruction. **The submandibular gland has a poor capacity for recovery following infection**. Despite control of acute symptoms with antibiotics, the gland becomes chronically inflamed.
- Calculi (80% of them occur in the submandibular salivary gland) commonly occur in the duct and also within the gland and produce recurrent sialoadenitis. Calculi are more common in the submandibular salivary gland than in the parotid gland because of the following reasons:
  1. **Higher mucin content** in the submandibular salivary gland secretions.
  2. **Calcium and phosphate** content in the secretion is high. Hence, 80% of them are radiopaque and are detected by plain X-ray (Fig. 18.6).
  3. **Nondependent drainage** of the secretions. Gland is in the neck and the duct opens into the oral cavity.
  4. **Kinking or hooking** of submandibular duct by lingual nerve.

Fig. 18.6: Submandibular calculi in the duct—removed (intraoral approach)

Clinical features (Key Box 18.5)

- **Salivary colic:** It is a severe pricking type of pain which is exaggerated at the time of meals. Salivary secretions are induced by a meal or lemon (Lemon juice test). As a result of blockage due to a stone, the tension within the gland increases, resulting in pain.
- **Lingual colic:** If a calculus is situated within the submandibular duct where it is hooked by lingual nerve, the

KEY BOX 18.5

SUBMANDIBULAR SALIVARY GLAND ENLARGEMENT

- Location—submandibular region
- Lobular, firm swelling
- Bidigitally palpable
- Stone may be palpable within the duct, intraorally

Fig. 18.7: Submandibular sialoadenitis

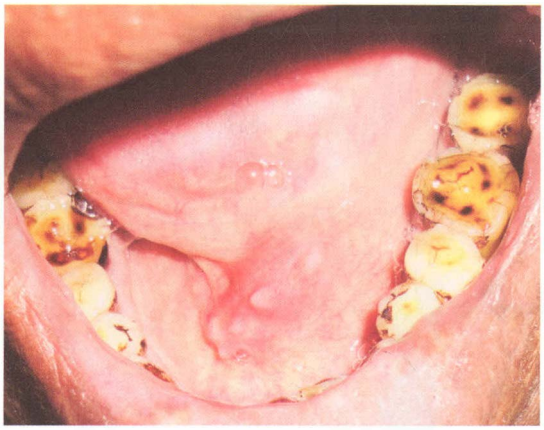


Fig. 18.8: Inflamed opening of submandibular duct

pain can radiate to the tongue as a result of irritation to the lingual nerve.

- **Enlargement of salivary gland during meals is the characteristic feature of salivary calculus**. Classically submandibular salivary gland swelling is located in the submandibular region. It is firm in consistency with a lobular surface. It is tender and both lobes are enlarged. **It is bidigitally palpable (submandibular lymph node is palpable only in the neck)** both inside the oral cavity and in the neck. The swelling reduces in size once the stimuli are withdrawn (after meals).
- The stone may be palpable within the gland (in the neck), within the duct (intraorally), or sometimes it may be seen at the orifice of the submandibular duct on the side of lingual frenulum.
- It is not uncommon to get a severe septic sialoadenitis with gross swelling of the gland and inflammatory oedema almost like Ludwig's angina (Fig. 18.7).

Treatment

- An oblique lateral or posterior oblique occlusal radiography may demonstrate a stone.
  1. **Stone in the submandibular duct:** This can be removed by incising the mucosa over the floor of the mouth, after stabilising the stone. Removal of the stone is followed by **gush of old dirty contents** of the submandibular gland (Fig. 18.8).
  2. **Chronic sialoadenitis:** This requires excision of submandibular salivary gland. Three steps of dissection of the gland include incision, mobilisation and excision (**Manipal Rule of 2**—Table 18.2).
- **Incision:** It should be a skin crease incision over the lower pole of the gland, the posterior limit of the incision should be at least 2 cm away from the angle of the mandible, to avoid damage to the cervical branch of facial nerve. The incision is deepened till the deep fascia is opened.
- **Mobilisation of the gland:** Division of the facial artery twice, once in a deeper plane on the posterolateral aspect and another at the superolateral aspect close to the lower border of the mandible is an important step which permits mobilisation of the gland. Separation of the gland from fibres of mylohyoid muscle by dividing small arteries completes the mobilisation (Fig. 18.9).
- **Excision of the gland:** It is done by ligating and dividing submandibular duct.

Complications

- Damage to lingual nerve, marginal mandibular nerve or even to hypoglossal nerve. Seroma and infection are the other complications.

Table 18.2 Submandibular salivary gland excision—**Manipal Rule of 2**

• 2 common indications	→	Stone and as a part of radical neck dissection
• 2" long incision	→	Curved incision over the swelling
• Protect 2 superficial nerves	→	Cervical and marginal mandibular branches of facial nerve
• Protect 2 deep nerves	→	Lingual and hypoglossal nerve
• Ligate facial artery 2 times	→	First at deeper plane and then at superficial plane
• Divide 2 muscles	→	Superficial—platysma; Deep—fibres of mylohyoid
• Remove 2 lobes	→	Superficial and deep
• Incision is 2 cm medial to mandible, 2 cm anterior to angle of the mandible	→	To protect 2 superficial nerves

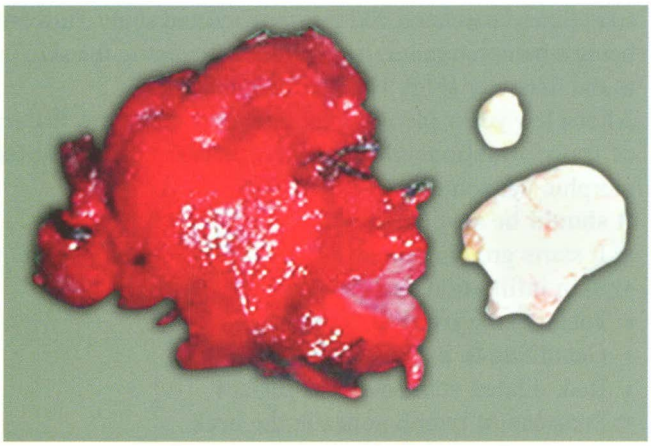


Fig. 18.9: Submandibular stone and gland

- Transection of the nerve to mylohyoid muscle produces **anaesthesia of the submental skin.**

SALIVARY GLAND TUMOURS

INTERNATIONAL CLASSIFICATION

I. Epithelial tumours
II. Nonepithelial tumours.

I. Epithelial tumours

A. Adenoma
   1. Pleomorphic adenoma
   2. Monomorphic adenomas
      - Adenolymphoma (Warthin's tumour)
      - Oxyphilic adenoma (oncocytoma)
      - Other types

B. Mucoepidermoid tumours
C. Acinic cell tumour
D. Carcinoma
   1. Carcinoma in pleomorphic adenoma
   2. Adenoid cystic carcinoma
   3. Undifferentiated carcinoma
   4. Adenocarcinoma
   5. Epidermoid carcinoma
   6. Acinic cell tumour
   7. Mucoepidermoid carcinoma
   8. Malignant mixed tumour.

II. Nonepithelial tumours

1. Lipoma
2. Lymphoma
3. Neurofibroma
4. Lymphangioma
5. Sarcoma

Salivary gland tumours are not uncommon. There are dozens of histological types of salivary gland tumours. However, pleomorphic adenoma and adenolymphoma are the common benign types. Carcinoma arising in pleomorphic adenoma, mucoepidermoid tumours and adenoid cystic carcinoma are important malignant tumours.

Incidence: 80% of salivary gland tumours are found in the parotid gland. Out of these, 80% are benign, of which 80% are pleomorphic adenomas.

In the submandibular salivary gland, 50% are benign and 50% are malignant.

In the minor salivary glands, 90% are malignant. Thus, the incidence of malignancy increases from major to minor salivary glands.

PLEOMORPHIC ADENOMA OF PAROTID GLAND (MIXED TUMOUR)

It is the most common benign salivary gland neoplasm.

Pathology

- Epithelial cells proliferate in strands, or may be arranged in the form of acini or cords.
- There are also myoepithelial cells which proliferate in sheets. They are called spindle-shaped cells.
- The tumour produces mucoid material, which displaces and separates the cells resembling cartilage in histological section.

PEARLS OF WISDOM

Because of the presence of epithelial cells, myoepithelial cells, mucoid material, pseudocartilage and lymphoid tissue, the tumour is called pleomorphic adenoma.

- As the tumour grows, it compresses the normal parotid tissue and the branches of the tumour penetrate the thin capsule and enter the substance of the parotid. Simple enucleation will result in a recurrence. Hence, superficial parotidectomy has to be done.

Clinical features

1. Middle-aged women, around 40 years, are commonly affected (Fig. 18.10) (female, fifth decade and fullness near ear lobule).

Fig. 18.10: Pleomorphic adenoma—classical signs

2. Typically, a history of a very slow growing swelling (for a few years) is usually present.
3. The swelling is painless. **Any painless swelling near the ear is best assumed to be parotid gland neoplasm unless proved otherwise** (Fig. 18.11).

Signs (Key Box 18.6)

1. Parotid swelling has the following classical features:
   - It presents as a **swelling in front**, below and behind ear.
   - **Raises ear lobule.**
   - **Retromandibular groove** is obliterated.
2. It is rubbery or firm. Soft areas indicate necrosis. In long-standing cases, it can be hard. Surface can be nodular or sometimes bosselated. Skin is stretched and shiny. Howeve being a benign tumour, it is neither adherent to the skin n to the masseter (Figs 18.12, 18.13 and 18.17).
3. After a few years, pleomorphic adenoma may show featur of transformation into malignancy (carcinoma explec morphic adenoma).

It should be suspected when

- It starts growing rapidly
- Skin infiltration occurs
- Facial nerve paralysis occurs
- Gets fixed to masseter muscle
- Red, dilated veins over the surface
- Presence of lymph nodes in the neck
- Tumour feels stony hard.

KEY BOX 18.6

CLINICAL EXAMINATION OF PAROTID TUMOURS

- Swelling firm, nodular
- Facial nerve involvement (80–90% cases of malignancy)
- Fixity to mandible and masseter
- Deep cervical nodes
- Opening of parotid duct
- Shift of tonsil and pillar of the fauces
- Other salivary glands (both sides).

INTRAORAL EXAMINATION (Fig. 18.14)

- Approximately 10% of the parotid tumours are behind th facial nerve in the deep lobe.
- This is appreciated by intraoral examination wherein th tumour presents with a parapharyngeal mass whic displaces the tonsil or soft palate medially.
- Deep lobe tumours present as dysphagia. Such tumours ma not show gross swelling on the outer aspect but as the grow, they pass through the stylomandibular tunnel of Pate

Fig. 18.11: Gross enlargement of parotid gland of 30 years duration

Fig. 18.12: Lateral view showing nodular surface

Fig. 18.13: Bosselated surface. Ear lobule is raised

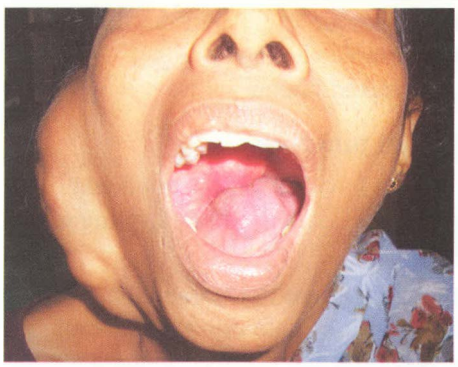


Fig. 18.14: Deep lobe tumour. Importance of intraoral examination

(*Courtesy:* Dr Sreejayan, Prof of Surgery, Calicut Medical College, Calicut, Kerala)

Fig. 18.15: Large sebaceous cyst in the preparotid region. It had sebaceous punctum

Fig. 18.16: Parotid cyst in a young boy—It was fluctuant and transillumination-negative. It was a haemangioma

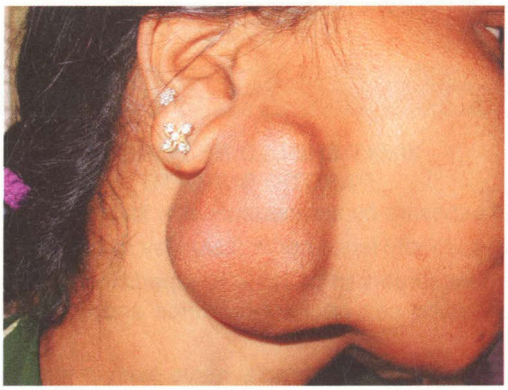


Fig. 18.17: Nodular surface of pleomorphic adenoma

KEY BOX 18.7

DIFFERENTIAL DIAGNOSIS OF SWELLING IN THE PAROTID REGION

1. **When it involves the preparotid region and angle of mandible**
   - Preparotid lymph node enlargement: It may be enlarged due to tuberculosis, metastasis or in non-Hodgkin's lymphoma. Sebaceous cyst in pre-parotid region (Fig. 18.15)
   - Preauricular dermoid cyst (young patients)
   - Mesenchymal tumours such as lipoma, neurofibroma
   - Haemangioma, lymphangioma (Fig 18.16)
2. **When it involves lower pole of parotid**
   - Upper jugular chain lymph node enlargement—metastatic or tubercular
   - Branchial cleft cyst
   - Epithelial inclusion cyst

and push the pharyngeal wall, tonsil and soft palate. These tumours are called **Dumbbell tumours** (Key Box 18.7)

DIFFERENTIAL DIAGNOSIS OF SWELLING IN PAROTID REGION

Investigations

Slow growing parotid tumours **should not be subjected to biopsy** for 2 reasons:

- **Injury to the facial nerve**
- **Seeding of tumour** cells in the subcutaneous plane which causes recurrence in about 40–50% of cases.

1. **Fine needle aspiration cytology** (FNAC) is done to confirm the diagnosis and rule out malignancy.
2. **CT scan** is done when the tumour is arising from the **deep lobe**. It helps to define the **extraglandular spread**, the extent of parapharyngeal disease, cervical lymph nodes and bony infiltration.
3. **FNAC of the lymph nodes** that are palpable in the neck in cases of malignancy of the parotid gland.
4. **X-ray of the bones** (mandible and mastoid process) to look for bony resorption, if malignancy is suspected.
5. **MRI is a better investigation. However, it is expensive—CT Scan and MRI lack specificity for differentiating between benign and malignant lesions.**

Indications for CT scan

1. Suspected bone destruction at skull base
2. Suspected involvement of mandible
3. To assess neck nodes.

Indications for MRI

1. Delineating the interface between tumour and normal salivary gland
2. Better imaging of the parapharyngeal space
3. Evaluating perineural spread, e.g. adenoid cystic carcinoma
4. Facial nerve status may be better appreciated.

Treatment

- Conservative superficial parotidectomy (Fig. 18.18 and Key Box 18.8)
- It is the standard surgery done for benign pleomorphic adenoma. It means removal of the entire lobe containing the tumour which is superficial to the facial nerve. **Facial nerve should always be preserved**. Enucleation should never be done as it causes recurrence and can injure facial nerve. It is difficult to remove a recurrent tumour.

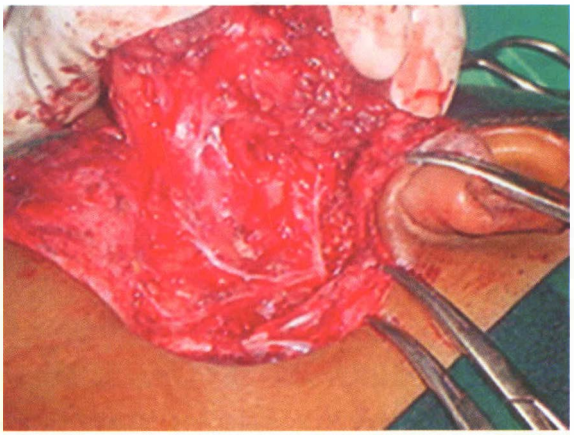


Fig. 18.18: Conservative superficial parotidectomy: Facial nerve divides the gland into superficial and deep lobes. The entire superficial lobe is removed preserving the facial nerve

KEY BOX 18.8

CONSERVATIVE SUPERFICIAL PAROTIDECTOMY

- Indicated in pleomorphic adenoma and other benign neoplasms
- Tumour along with the normal lobe is removed
- Preserve the facial nerve, **even in malignant tumours unless grossly involved.**
- Avoid rupture of the gland (Fig. 18.19)
- **Enucleation should not be done as it causes recurrence.**

Some important steps of superficial parotidectomy (Key Box 18.9)

1. **Adequate exposure** by an incision which starts in front of tragus of ear, vertically descends downwards, curves round the ear lobule up to the mastoid process and is carried downwards in the neck ('Lazy S' incision).
2. **Recognising the facial nerve at surgery** (Fig. 18.19)
   - Facial nerve lies **1 cm inferomedial to the bony cartilagenous junction of external auditory canal (Conley's pointer).**

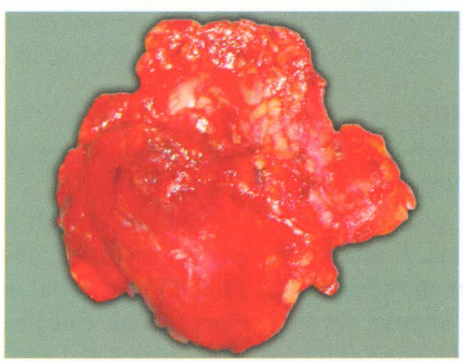


Fig. 18.19: Superficial parotidectomy specimen

   - **Trace the posterior belly of digastric** up to the mastoid process. Facial nerve is in between the muscle and tympanic plate.
   - A **nerve stimulator** may be used.
3. **Developing a plane:** Facial nerve and retromandibular vein divide the parotid gland into superficial and deep lobes. Benign tumours do not invade this **faciovenous plane of Patey**.
4. **Gentle handling**, good suction and perfect haemostasis help in clear recognition of the nerve.
5. ***In toto* removal**—The tumour along with **the lobe** to avoid spillage (which is one of the causes of recurrence).
6. **Good suction drainage** of the wound is necessary to avoid haematoma, wound infection, etc.

KEY BOX 18.9

WHAT SHOULD BE DONE IN PAROTID SURGERY

- Wide exposure by an adequate skin flap
- Always identify facial nerve (best way to avoid injury).
- Minimum surgery to be done is superficial parotidectomy (enucleation can cause recurrence and injury to facial nerve)
- Always try to preserve facial nerve even in malignancies unless it is directly infiltrated.
- If facial nerve is excised, try to reconstruct immediately by nerve graft—greater auricular or sural nerve.
- Always 'drain' the cavity.

Total conservative parotidectomy excision of superficial and deep lobe of parotid gland while preserving the facial nerve.

Total parotidectomy with the excision of facial nerve when nerve is involved.

Radical parotidectomy involves excision of other structures along with parotid gland and facial nerve

- Skin
- Infratemporal fossa
- Mandible
- TM joint
- Petrous bone

Indications for postoperative radiotherapy

- If the deep lobe is involved
- If the lymph nodes are involved
- High grade tumours
- If margins are positive
- Perineural invasion
- Lymphovascular invasion

Postoperative radiotherapy

- T3/T4 cancer: Adenoid cystic carcinoma recurrent tumours

ADENOLYMPHOMA (WARTHIN'S TUMOUR, PAPILLARY CYSTADENOMA LYMPHOMATOSUM)

- Adenolymphoma is not a lymphoma. It is a misnomer *(vide infra)*.
- It is a benign parotid tumour and next common to pleomorphic adenoma. It constitutes about 10% of parotid tumours.
- **Origin of adenolymphoma:** During development some parotid tissues get included within lymph nodes (preparotid) which are present within the parotid sheath.

Histology

- It is composed of double layered eosinophilic epithelium. **The inner cells are columnar**.
- Presence of lymphatic tissue in the stroma, and lymph follicles is characteristic of adenolymphoma (hence the name).

Clinical features (Table 18.3)

- Middle aged or elderly males are commonly affected – usually **they are smokers**.
- Can be **bilateral**, in some cases (10%).
- It has smooth surface, round border with soft, cystic consistency (Fig. 18.20).
- Classically, situated at the lower pole of parotid elevating the ear lobule. Sometimes it may be **multicentric**.
- **This tumour affects only parotid gland** (very, very rarely other glands may be affected).

Treatment

- It has got a well-defined capsule. Hence, enucleation used to be done earlier **but not now. Superficial parotidectomy is the treatment of choice.**

Table 18.3 Comparison between pleomorphic adenoma and adenolymphoma

Features	Pleomorphic adenoma	Adenolymphoma
1. Incidence	70–80%	10%
2. Sex	Common in females	Common in males
3. Number	Single	Sometimes multiple
4. Site	Unilateral	Bilateral
5. Clinical feature	Nodular, firm	Smooth, soft cystic
6. Histology	Pleomorphism	Double layer epithelium and lymphoid tissue
7. ^{99m}Tc-Pertechnetate scan	Cold spot	Hot spot
8. Treatment	Superficial parotidectomy	Superficial parotidectomy

MUCOEPIDERMOID TUMOUR

- As the name itself suggests, it consists of sheets of epidermoid cells and cystic spaces lined by mucus secreting cells (Fig. 18.21).
- **In childhood, it is the commonest parotid tumour.** They are benign, slow-growing but hard in consistency. (Adenolymphoma and mixed tumours are firm but mucoepidermoid tumour is hard). Parotid is the commonest site. In cases of minor salivary glands, palate is the commonest site.
- Mucoepidermoid tumours can infiltrate local tissues, lymph nodes or skin. Hence, a few consider that **mucoepidermoid tumours are always carcinomatous.**
- **Well-differentiated** tumours behave like benign tumours, **intermediate ones** are aggressive and **undifferentiated** tumours metastasise early. Mucoepidermoid carcinoma is the most common malignant epithelial neoplasm of salivary gland.
- The **low grade** tumours are composed of predominantly **mucus secreting cells**. **High grade tumours** have predominantly **epidermoid cells**.
- Benign tumours need excision and malignant tumours need radical parotidectomy. Radiation is required in the post-operative period.

OTHER TUMOURS

Acinic cell tumour

- These are the uncommon parotid tumours. Commonly occur in women.
- The cells resemble those of serous acini and this tumour also has properties of invasion such as mucoepidermoid tumour. It tends to be soft and sometimes cystic.

Oxyphil adenoma

- Also called oncocytoma. It occurs exclusively in the parotid gland. It is a solid tumour and occurs in the sixth decade of life.

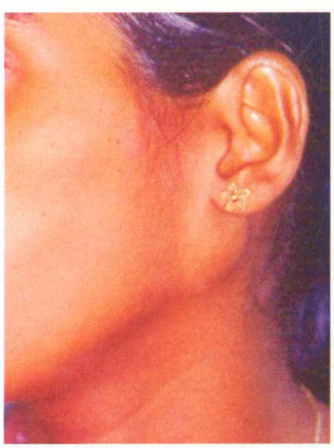


Fig. 18.20: Adenolymphoma

Fig. 18.21: Mucoepidermoid carcinoma

Adenoid cystic carcinoma

- It is a highly malignant tumour consisting of cords of dark staining cells with cystic spaces containing mucin. It also consists of myoepithelial cells and duct epithelium.
- Even though **slow-growing**, it spreads along the **perineural tissue,** may invade periosteum or medullary bone at a distance. This bone resorption results in bony tenderness.
- These tumours have a **high incidence of distant metastasis** but in general **they display indolent growth**. Skip lesions are common as it spreads along the nerve tissue, which leads to treatment failure.
- Local infiltration, lymphatic and blood spread, and local recurrence are important features.
- It is **hard and fixed** and can produce **anaesthesia of the skin overlying the tumour.**
- Early cases are treated by radical parotidectomy with block dissection of the neck. However, many cases present late to the hospital. Thus, palliative radiotherapy is given to reduce pain and to arrest progress of the disease.

Complications of parotidectomy (Key Box 18.10)

KEY BOX 18.10

COMPLICATIONS OF PAROTIDECTOMY

- **Flap necrosis**—avoid acute bending of the incision and to use gentle retraction
- **Facial nerve palsy**—careful identification
- **Fluid collection**: Blood or seroma—perfect haemostasis and drain should be used
- **Fistula salivary**—duct should be ligated
- **Frey syndrome**—occurs in 10% of the cases

Observe **5 Fs.**

Summary of malignant salivary gland tumours (Figs 18.22 to 18.29 and Table 18.4)

To find out the exact type of malignant tumour is of interest to pathologists. Clinically, one can suspect malignancy when a salivary tumour has one of the following features:

- Rapidly growing neoplasm
- Change in consistency (the tumour tends to be hard)
- Fixity to underlying muscle such as masseter as in parotid tumours
- Fixity to mandible as in parotid or submandibular tumour
- Involvement of facial nerve as in 80% of cases of malignant parotid tumours.
- Resorption of adjacent bone such as mastoid, tenderness as in adenoid cystic carcinoma.
- Significant hard nodes in the neck.
- They are treated by radical sialadenectomy with radical block dissection of the neck. Radiotherapy is used as a palliative treatment.

'Most common' for salivary glands (Key Box 18.11)

KEY BOX 18.11

'MOST COMMON' FOR SALIVARY GLANDS

- Most common benign parotid tumour in adults—pleomorphic adenoma
- Most common benign parotid tumour in children—haemangioma
- Most common type of cancer arising in the parotid glands is mucoepidermoid cancer.
- Most common malignant tumour in submandibular gland—adenoid cystic carcinoma
- Most common minor salivary gland tumour is adeno-carcinoma
- Most common site of squamous cell carcinoma is sub-mandibular salivary gland
- Most common response to radiotherapy among the malignant tumours is adenoid cystic carcinoma.

Table 18.4 Comparison between submandibular and parotid tumours

	Submandibular tumour	Parotid tumour
• Incidence	Uncommon	Common
• Malignancy	50% of tumours	10–20%
• Location	Submandibular triangle	Parotid region
• Deep lobe involvement	Intraorally felt in the floor of mouth	Tonsillar shift, can also be felt in the lateral wall
• Surgical treatment of malignant tumour	Small tumours—intracapsular submandibular excision Large tumours—radical excision with or without sacrifice of '2' nerves—lingual and hypoglossal	Total conservative parotidectomy for malignant tumours. Facial nerve should always be preserved.
• Lymph nodes	Selective neck dissection—supraomohyoid neck dissection	Selective neck dissection (lateral) levels II, III, IV and V lymph nodes
• **Warthin's**	**Does not occur here**	**Exclusively occurs in the parotid gland.**

MALIGNANT PAROTID TUMOURS

Fig. 18.22: Low grade mucoepidermoid carcinoma—had restricted mobility due to fixity to masseter

Fig. 18.23: Adenoid cystic carcinoma—perineural spread is common with this tumour

Fig. 18.24: High grade mucoepidermoid carcinoma—rapidly growing and dilated veins over the surface

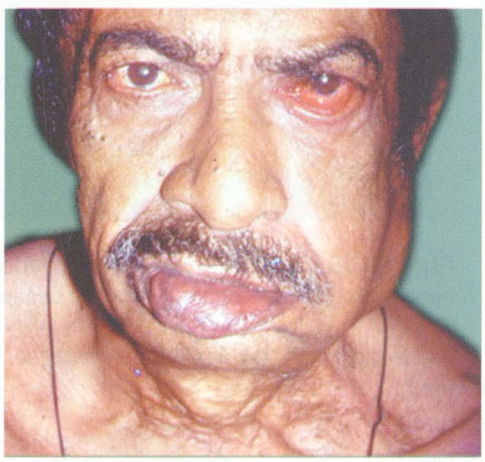


Fig. 18.25: Facial nerve paralysis—one of the strong clinical signs of malignancy in a parotid tumour

Fig. 18.26: Adenoid cystic carcinoma—hard and fixed receiving radiotherapy

Fig. 18.27: Malignant mixed tumour. Ulcerated, late cases

Fig. 18.28: Carcinoma parotid—late stage with involvement of platysma—**platysma sign**

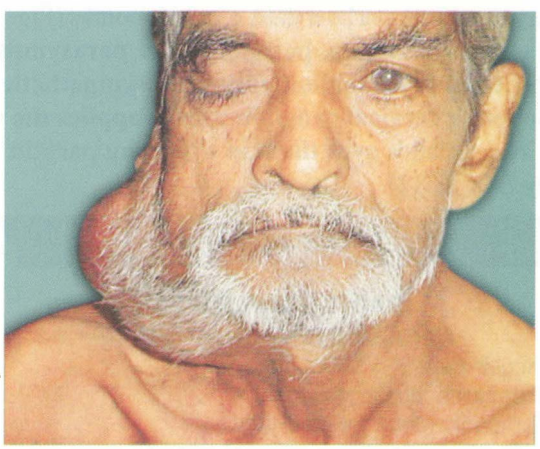


Fig. 18.29: Same patient in Fig. 18.28 also had facial nerve paralysis

FEW OPERATIVE PHOTOGRAPHS (Figs 18.30 to 18.35)

Fig. 18.30: Malignant mixed tumour—position of patient

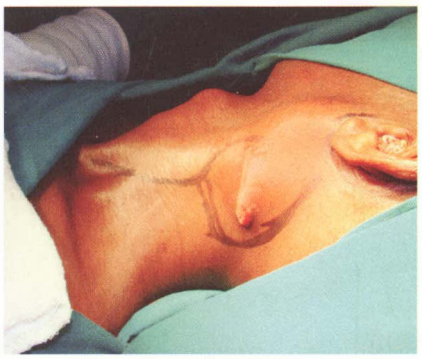


Fig. 18.31: Methylene blue is used to mark the site of incision and part to be raised

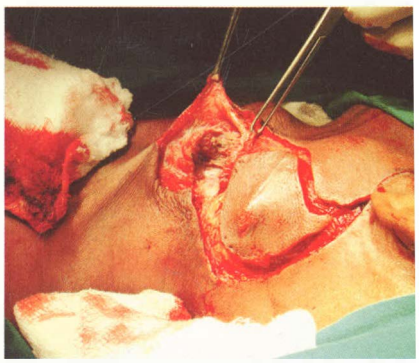


Fig. 18.32: Flaps are elevated

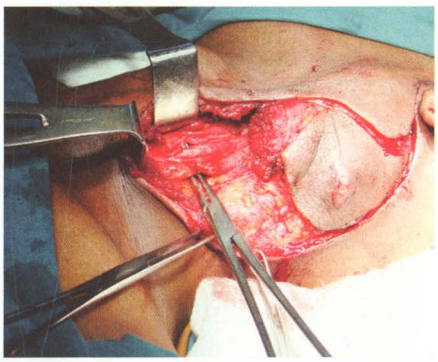


Fig. 18.33: Strap muscles are divided as a part of supraomohyoid block dissection

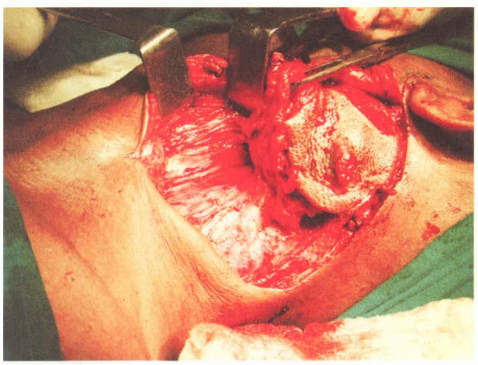


Fig. 18.34: Wide excision and block dissection

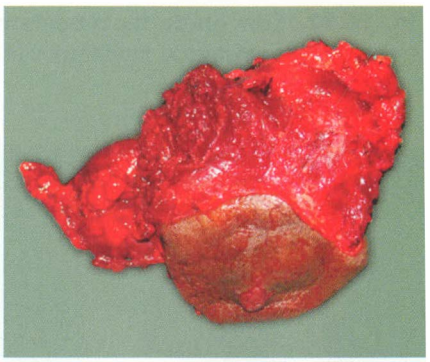


Fig. 18.35: Specimen of tumour with surrounding fat, fascia and lymphatic tissue

MISCELLANEOUS

FREY'S SYNDROME—GUSTATORY SWEATING

- It occurs after surgery for parotid tumours, surgery in the region of temporomandibular joint or due to injury to the parotid gland. Injury to the auriculotemporal nerve can occur at a site where it turns around the neck of the mandible. The injury manifests at a later date, e.g. 2–3 months (Fig. 18.36).
- Because of the injury, **postganglionic parasympathetic fibres from otic ganglion unite with sympathetic fibres of superior cervical ganglion** which **supplies the vessels** and sweat glands **over the skin overlying parotid region** (Key Box 18.12).

KEY BOX 18.12

PARTS SUPPLIED BY AURICULOTEMPORAL NERVE

- Auricular part:
  - External acoustic meatus
  - Tympanic membrane surface
  - Skin of auricle above external acoustic meatus
- Temporal part: Hairy skin of the temple.

- As a result of this, whenever the **act of chewing** or **mastication is started,** there is increased sweating and hyperaesthesia in the region supplied by auriculotemporal nerve **(cutaneous branch of mandibular division of trigeminal nerve).** Hence, it is called **auriculotemporal syndrome**.

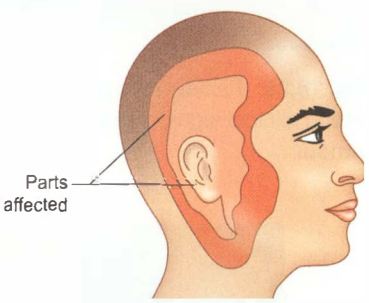


Fig. 18.36: Frey's syndrome

- **Diagnosis: Starch iodine test**—paint the affected area with iodine and allowed it to dry before applying the dry starch.
- The starch turns blue on exposure to iodine in the presence of sweat.

Prevention

- Principle is to provide a barrier between the skin and parotid bed by using temporalis fascial flap or sternomastoid muscle flap.

Treatment

1. **Reassurance, aluminium chloride—antiperspirant** which is a useful astringent
2. Denervation by **tympanic neurectomy**
3. Latest treatment includes injection of **botulinum toxin into the affected skin** (*see* page 441 also).

RARE CAUSES OF SALIVARY GLAND ENLARGEMENT

1. Sjögren's syndrome

It is the diffuse infiltration of salivary and lacrimal glands with lymphocytes resulting in enlargement of glands and slow destruction of acini. Thus, clinical features include dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia). These along with a third component rheumatoid arthritis, form the triad of **Sjögren's syndrome (primary)**.

- 30% of patients with systemic lupus erythematosus and all patients with primary biliary cirrhosis develop Sjögren's syndrome. This is termed **secondary Sjögren's syndrome**.
- Other features: This disease is 10 times more common in females and presents with painful enlargement of the glands.
- **Complications**
  1. Lymphomatous transformation (high in primary).
  2. Oral candidiasis.

2. Mikulicz disease

Due to autoimmune mechanism, symmetrical enlargement of all salivary glands and lacrimal gland enlargement occur. Dry mouth and narrow palpebral fissures are diagnostic of this condition.

3. Drugs

Carbimazole and **thiouracil** can cause enlargement of salivary glands.

4. Metabolic disorders

Diabetes and **acromegaly** are the other causes.

5. Granulomatous sialoadenitis

These are rare, painless swellings. Following are the causes:

- **Tuberculosis**
- **Sarcoidosis**—commonly affects parotid gland wherein it is called **pseudotumour**
- **Toxoplasmosis**
- **Cat-scratch** disease
- **Wegener's granulomatosis**

PAROTID FISTULA

It is an uncommon condition which commonly occurs after surgery on the parotid gland (Key Box 18.13).

KEY BOX 18.13

PAROTID FISTULA

- Any surgery on the parotid gland—superficial parotidectomy, drainage of abscess, surgery for carcinoma cheek, facio-maxillary trauma are the causes.
- Discharging watery fluid, exaggerated by keeping lime in the mouth.
- Fistulogram confirms the diagnosis.
- Exploration and excision of fistula and ligation of duct is required.

MINOR SALIVARY GLAND TUMOUR (Fig. 18.37)

- Even though they are called minor, numberwise they are major (many), about 450 in number.
- They are mucus-secreting.
- They can present as mucus retention cyst (common in lip) or as malignant tumour.
- 90% of minor salivary gland tumours is malignant.
- Since they are submucosal, they start as a submucous nodule (very important point in the history) to differentiate from carcinoma buccal mucosa/lip, etc.
- As they grow, they ulcerate. Ulceration is a feature of malignancy.
- Slowly lymph nodes get enlarged.
- Treatment of benign cyst/tumour is by simple excision and malignant tumour is by wide excision.

Fig. 18.37: Minor salivary gland tumour in the palate. Laser excision was done

SURGERY FOR FACIAL NERVE PALSY

Indications for different types of surgery

1. Early immediate nerve repair, in case of injury to the nerve.
2. **Late nerve crossing** by suturing peripheral branches of facial nerve to one of the following nerves:
   - Hypoglossal nerve, spinal accessory nerve
   - Phrenic nerve.
3. Surgery to achieve movement in long standing facial palsy (usually after 1 year).

A. Static procedures

- Suspension of lips, cheek and angle of mouth to zygomatic bone or temporal fascia using fascia lata, palmaris longus tendon or other alloplastic materials.
- Medial canthoplasty to reduce epiphora
- Lateral tarsorrhaphy (canthoplasty) to prevent exposure keratitis due to widened palpebral fissure.

B. Dynamic procedures

- Muscle transfer with carefully preserved muscle nerve and vessel, e.g. temporalis muscle transfer, masseter muscle transfer.
- Cross face nerve transplantation using sural nerve. Using microscope, sural nerve is sutured to the two or three relatively insignificant branches of facial nerve (selected by intraoperative electric stimulation) on normal side. Other end of the sural nerve is sutured to distal end of the divided facial nerve on paralysed side.
- Free neurovascular gracilis muscle graft using microvascular techniques.

PERIPHERAL NERVE REPAIR AND TRANSFERS

PRINCIPLES OF NERVE REPAIR

1. Preoperative assessment of motor and sensory system
2. Microsurgical tools should be available
3. Tension free repair should be done. If tension free repair is not possible, then nerve graft can be used
4. Primary repair when conditions permit
5. Delayed repair should be done in case of extensive injury.

Microsurgical techniques

- 4X magnification needed
- 9–0/10–0 nylon is used for repair
- Ends prepared with microscissors. First suture to be loose to ease the alignment of other sutures.
- Avoid postural manoeuvre to avoid tension. Nerve can be mobilised 1–2 cm proximally and distally for tension free suturing but not more than that.

Conduits

- To place a polyglycolic acid tube to bridge the gap rather than to perform nerve grafting.
- **Indication**
  - Donor nerve harvest not possible due to availability and associated morbidity
  - Patients who decline autogenous nerve graft harvest
  - Nerve gap of not more than 3 cm.
- **Principle of conduits**
  - Nerve regeneration and proximal and distal axonal matching result from a combination of neurotropism and contact guidance.

TYPES

I. Nerve grafting

- **Indication:**
  - If a tension free repair is not possible
  - If the nerve gap is more than 3 cm
  - Can be used in place of conduits
- **Principle**
  - The graft should be oriented in a reverse fashion from its native position so that the regenerating fibres are not diverted from the distal neurorrhaphy site and distal stump. Misalignment should be avoided.
- If the nerve gap is small, fascicular matching can be done
- If the nerve gap is long, techniques of awake stimulation or histochemical staining may be used.
- **Donor sources for nerve grafting**
  - Posterior interosseous nerve, medial antebrachial cutaneous nerve for small digital nerves, sural nerve for nerve gaps involving larger nerves, greater auricular nerve.

II. Nerve transfer

- It is used for reconstruction of proximal nerve injuries.
- **Drawbacks** of nerve graft that become the merits of nerve transfer:
  - Grafting across a proximal nerve injury has poor functional outcomes
  - Timely re-innervation may not be possible if the target muscle is too far from the regenerating site.
- A nerve transfer converts a high level nerve injury to a low level nerve injury by recruiting expendable nerve fascicles from the donor nerve to innervate critical nerves close to their target end organs.
- Donor nerves are preferentially selected according to their proximity to the target site
- **Criteria for nerve transfers**
  - An expendable donor nerve
  - Donor nerve with a large number of pure axons
  - Donor nerve near the target organ
  - Donor motor nerve that innervates a muscle that is synergistic to the target muscle
- Nerve transfers are done for both motor and sensory nerves.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated.
- A few photographs and key boxes have been added.
- Granulomatous sialoadenitis, peripheral nerve repair and transfers have been added.

MULTIPLE CHOICE QUESTIONS

1. Commonest tumour of minor salivary gland is:
A. Pleomorphic adenoma
B. Warthim's tumour
C. Malignant tumour
D. Mucoepidermoid tumour

2. Parotid duct opens opposite:
A. Upper canine tooth
B. Lower canine tooth
C. Upper 2nd molar tooth
D. Lower 2nd molar tooth

3. Chronic parotitis in children is pathognomonic of which infection?
A. HCV infection
B. HBV infection
C. HIV infection
D. Syphilis

4. Which is the common site of calculi in submandibular salivary gland?
A. Superficial lobe
B. Deep lobe
C. Accessory lobe
D. Duct

5. Deep lobe tumours of parotid presents as following features *except:*
A. Dysphagia
B. Can push tonsils
C. Can push soft palate
D. Can block the external auditory meatus

6. The investigation of choice in deep lobe tumours of parotid gland is:
A. CT scan
B. Angiography
C. MRI
D. Ultrasound

7. Conley's pointer refers to:
A. Location of facial nerve in relation to tragal cartilage
B. Location of facial nerve in relation to posterior belly of diagstric
C. Location of facial nerve in relation to retromandibular vein
D. Location of facial nerve in relation to pinna

8. Following are true for chronic hyperplastic candidiasis *except:*
A. Invasion of *Candida albicans*
B. Antifungal treatment helps
C. High malignant potential
D. Floor of the mouth is affected

9. Following benign tumours have high incidence of recurrence *except*?
A. Adamantinoma
B. Deep lobe tumours of parotid gland
C. Desmoid tumours
D. Diffuse lipomata

10. Recommended treatment of pleomorphic adenoma is:
A. Enucleation
B. Excision
C. Superficial parotidectomy
D. Wide excision

11. Which benign tumour of parotid produces Hot Spot in Technetium scan?
A. Pleomorphic adenoma
B. Adenoid cystic carcinoma
C. Mucoepidermoid tumour
D. Adenolymphoma

12. Commonest parotid tumour in children is:
A. Pleomorphic adenoma
B. Warthim's tumour
C. Mucoepidermoid tumour
D. Lymphangioma

13. Most common malignant tumour in submandibular salivary gland is:
A. Pleomorphic adenocarcinoma
B. Adenoid cystic carcinoma
C. Mucoepidermoid tumour high grade
D. Acinic cell tumour

14. Following are true for precancerous lesions of lip/ oral cavity *except:*
A. Keratoacanthoma
B. Leukoplakia
C. Erythroplakia
D. Submucous fibrosis

15. Following are features of carcinoma maxillary antrum *except:*
A. Can cause asymmetry of face
B. Can cause proptosis
C. Can cause infraorbital nerve paralysis
D. Can cause buccal branch of facial nerve paralysis

16. Following are the boundaries of nasopharyngeal space *except:*
A. Nasal fossae
B. Basilar part of occipital bone
C. Body of sphenoid
D. Cribriform plate of ethymoid bone

17. Following are true for nasopharyngeal carcinoma *except:*
A. Presents as high anterior cervical lymphadenopathy
B. Can present as trigeminal neuralgia
C. Compression on IX and X cranial nerves
D. Can present as ophthalmoplegia

18. Following are painless ulcers in the tongue *except*:
A. Gummatous ulcers B. Carcinomatous ulcers
C. Systemic diseases D. Tuberculous ulcers

19. Following are painful ulcers in the tongue *except:*
A. Gummatous ulcers
B. Tuberculous ulcers
C. Aphthous ulcers
D. Dental ulcers

20. Following are true for syphilitic lesions of the tongue *except:*
A. Snail track ulcers B. Gumma
C. Hutchinson's wart D. Hunterian chancre

21. Following are true for dental cyst *except:*
A. Upper jaw is commonly involved
B. It is a large unilocular cyst
C. Cyst contains cholesterol crystals
D. Arises from unerupted tooth

22. Which one of the following swelling does not contai cholesterol crystals?
A. Branchial cyst
B. Sebaceous cyst
C. Dental cyst
D. Hydrocoele

23. Following are true for dentigerous cyst *except*:
A. Upper jaw is commonly involved
B. Produces egg shell crackling
C. X-ray shows soap bubble appearance
D. Arises from unerupted tooth

24. Adamantinoma of the jaw has following feature *except:*
A. It is a malignant tumour
B. It spreads within medullary bone
C. It is treated by wide excision
D. Mandible is the most common site

25. Which of the following is unilocular cyst?
A. Adamantinoma
B. Dentigerous cysts
C. Epididymal cyst
D. Dental cyst

ANSWERS

1 C	2 C	3 C	4 D	5 D	6 C	7 A	8 D	9 D	10 C
11 D	12 C	13 B	14 A	15 D	16 D	17 A	18 D	19 A	20 D
21 D	22 B	23 A	24 D	25 D					

19

Thyroid Gland

- Surgical anatomy
- Physiology
- Thyroid function tests
- Clinical examination
- Goitre
- Multinodular goitre
- Retrosternal goitre
- Graves' disease
- Malignant tumours
- Anaplastic carcinoma
- Solitary nodule
- Thyroiditis
- Lingual thyroid
- Ectopic thyroid

Introduction

Thyroid gland is an endocrinal gland present in the neck secreting T3 and T4 hormones. It is richly vascular and highly functional. Effects of hormonal changes affect every part of the body such as central nervous system, cardiovascular system, gastrointestinal system and reproductive system. It is also the site of various diseases—a simple enlargement, toxicity and malignant transformation.

SURGICAL ANATOMY OF THYROID GLAND

Development and Anatomy

- It develops from **median down growth** (midline diverticulum) of a column of cells from the pharyngeal floor between first and second pharyngeal pouches.
- The descent is anterior to structures that form hyoid bone and larynx (Fig. 19.1).
- By 6 weeks of intrauterine life, the **central column,** which becomes the thyroglossal duct, gets reabsorbed.
- The duct bifurcates to form thyroid lobes.
- **Pyramidal lobe** is formed by a portion of the duct.
- **Calcitonin** producing **parafollicular or C cells** originate from the fourth branchial pouch.
- Thyroid gland is present in the neck, enclosed by pretracheal fascia which is a part of deep cervical fascia. It has a right and left lobe joined by the **isthmus** in front of **2nd, 3rd and 4th tracheal rings**. It weighs about 20–25 g. A projection from the isthmus usually on the left side is called pyramidal lobe. It is attached to the hyoid bone by a fibrous band or muscle fibres called levator glandulae thyroideae.
- **Suspensory ligament of Berry:** This pair of strong condensed connective tissue binds the gland firmly to each side of cricoid cartilage and upper tracheal rings.

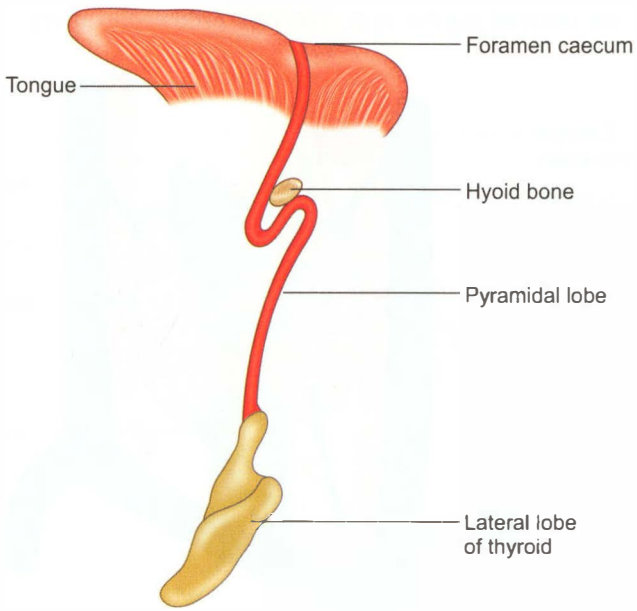


Fig. 19.1: Development of thyroid gland

- **Pretracheal fascia,** which is part of deep cervical fascia splits to invest the gland. These structures (ligament of Berry and pretracheal fascia) are responsible for thyroid gland moving with deglutition.

Arterial supply (Fig. 19.2)

- **Superior thyroid artery,** a branch of the external carotid artery, enters the upper pole of the gland, divides into anterior and posterior branches and anastomoses with ascending branch of inferior thyroid artery. Since the upper pole is narrow, ligation is easy.
- **Inferior thyroid artery** is a branch of thyrocervical trunk[1] and enters the posterior aspect of the gland. It supplies the gland by dividing into 4 to 5 branches which enter the gland at various levels (not truly lower pole).
- Inferior thyroid artery used to be ligated well away from the gland to avoid damage to RLN. However, ligation of these arteries on both sides will cause permanent hypoparathyroidism. Hence, *the current practice is to identify and ligate the branches of inferior thyroid artery (3–4) separately.*
- **Thyroidea ima artery** is a branch of either brachiocephalic trunk or direct branch of arch of aorta and enters the lower part of the isthmus in about 2 to 3% of the cases.

Venous drainage

- **Superior thyroid vein**[2] drains the upper pole and enters the internal jugular vein. The vein follows the artery.
- **Middle thyroid vein** is single[3], short and wide and drains into internal jugular vein.
- **Inferior thyroid veins** form a plexus which drain into innominate vein. They do not accompany the artery.
- **Kocher's vein** is rarely found (vein in between middle and inferior thyroid vein).

Nerves in relationship with thyroid gland

1. **Superior laryngeal nerve:** The vagus nerve gives rise to superior laryngeal nerve, which separates from it at skull base and divides into two branches. The larger **internal laryngeal nerve** is sensory to the supraglottic larynx. The smaller **external laryngeal nerve** runs close to the superior thyroid vessels and supplies cricothyroid.
   - This nerve is away from the vessels near the upper pole. Hence, during thyroidectomy, the upper pedicle should be ligated as close to the thyroid as possible.
2. **Recurrent laryngeal nerve (RLN)** is a branch of vagus, hooks around ligamentum arteriosum on the left and subclavian artery on the right and runs in the tracheoesophageal groove near the posteromedial surface. Close to the gland, the nerve lies in between (anterior or posterior) branches of inferior thyroid artery (Figs 19.3 and 19.4 in the Riddle's triangle).
   - On the right side, it is 1 cm within the tracheoesophageal groove (Key Boxes 19.1A and B).

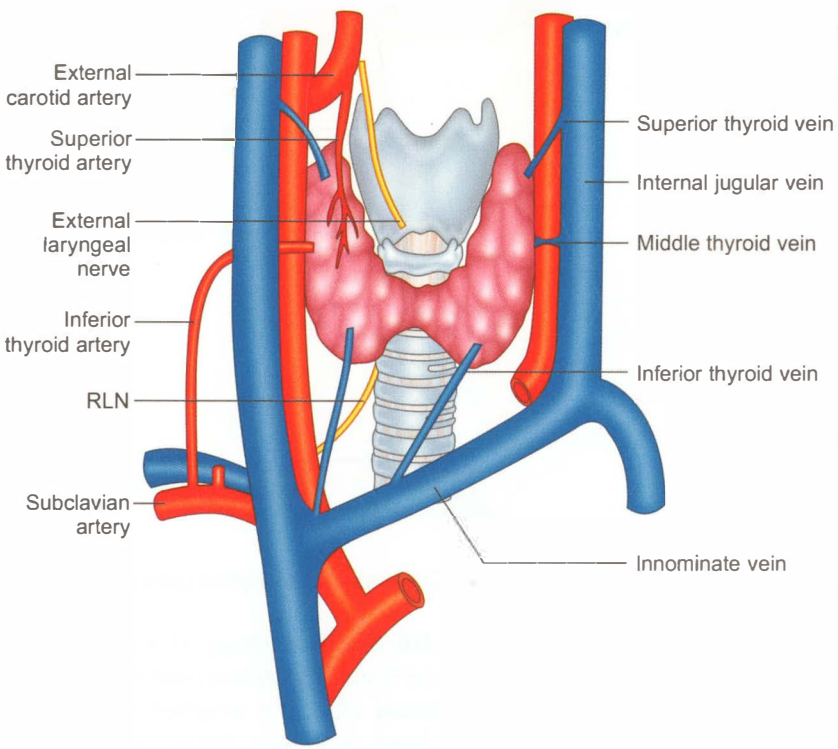


Fig. 19.2: Surgical anatomy of the thyroid gland

KEY BOX 19.1A

RECURRENT LARYNGEAL NERVE AT SURGERY

- Lack of colour—white in colour
- Lack of elasticity
- Lack of pulsation
- Longitudinal vein on the surface
- Longitudinal course
- Location—**Riddle's triangle**—it is between inferior thyroid artery superiorly, carotid artery laterally and trachea medially. From here RLN runs upwards to enter larynx at greater cornu of thyroid cartilage

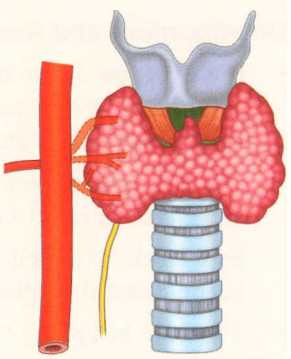


Fig. 19.3A

[1]Branches of thyrocervical trunk can be remembered as SIT—suprascapular, inferior thyroid and transverse cervical artery.
[2]Superior thyroid artery and vein are like newly married couple, they go together, hand in hand.
[3]Middle thyroid vein is single, a bachelor and inferior thyroid artery and vein are a divorced couple.

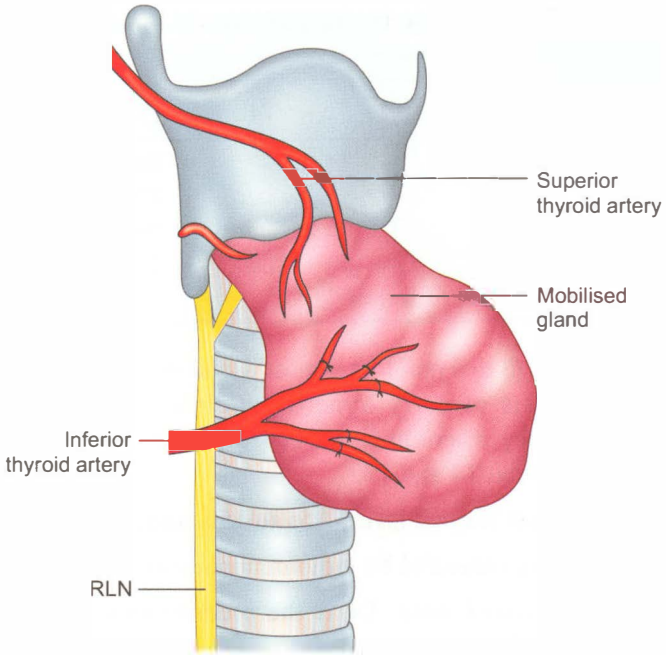


Fig. 19.3B: Course of recurrent laryngeal nerve—gland is mobilised. The branches of inferior thyroid are ligated (not the main trunk)

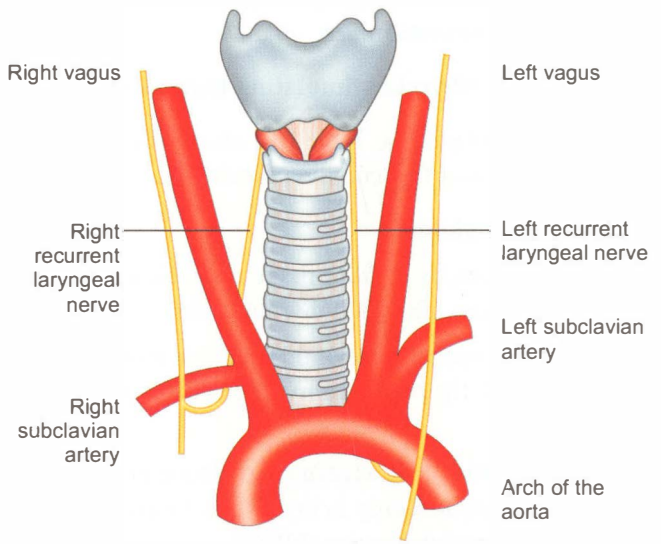


Fig. 19.4: Course of recurrent laryngeal nerve and origin

KEY BOX 19.1B

RECURRENT LARYNGEAL NERVE ANOMALIES

- The nerve traverses through the gland in about 5–8% of cases.
- The nerve may be very closely adherent to the posteromedial aspect of the gland.
- Nerve not seen—may be far away in the tracheo-oesophageal groove.
- Nonrecurrent, recurrent laryngeal nerve is found in about 1 in 1,000 cases. The nerve has a horizontal course.
- In 25% of the cases, it is within the ligament of Berry.

Lymphatic drainage of thyroid

- Subcapsular lymphatic plexus drains into pretracheal nodes (delphic nodes means uncertain) and prelaryngeal nodes which ultimately drain into lower deep cervical nodes and mediastinal nodes (Fig. 19.5).
- The chief lymph nodes are middle and lower deep cervical lymph nodes (Levels III and IV).
- Supraclavicular nodes and nodes in the posterior triangle can also be involved in malignancies of the thyroid gland, especially papillary carcinoma thyroid.

Histology (Fig. 19.6)

- Microscopically, it is divided into lobules.
- Each lobule has 20–40 follicles.
- Each follicle is lined by cuboidal epithelial cells.
- In the centre, colloid is present which is secreted from epithelial cells in response to calcitonin.
- Parafollicular cells are present in the interfollicular stroma.

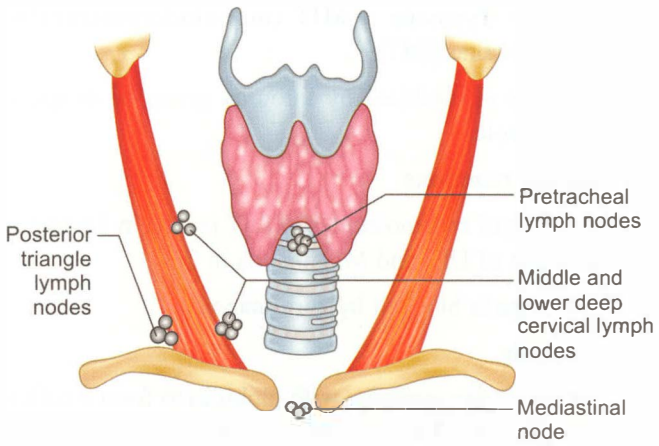


Fig. 19.5: Lymphatic drainage

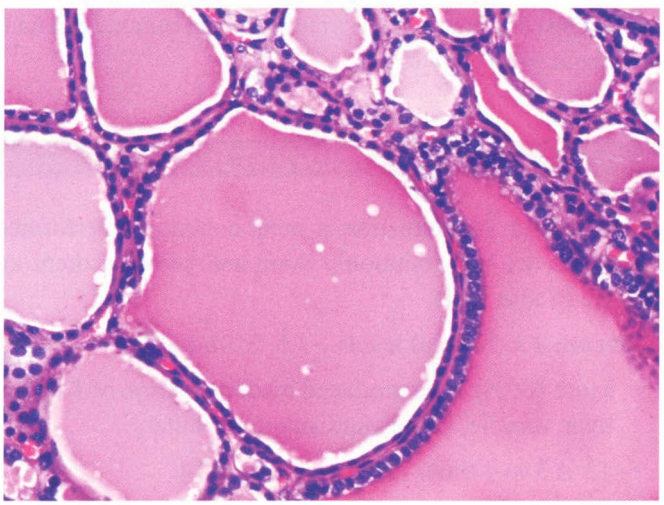


Fig. 19.6: Diffuse colloid goitre—Acini lined by cuboidal follicular cells with luminal colloid (*Courtesy:* Dr Laxmi Rao, HOD Pathology, KMC, Manipal)

PHYSIOLOGY

Tri-iodothyronine (T3) and thyroxine (T4) are the hormones secreted by the thyroid gland. Dietary requirement of iodine per day is 100–200 micrograms or 0.1 mg. Sources of iodine are milk, dairy products and sea food including fish.

Steps involved in the synthesis of these hormones

1. **Iodide trapping** from the blood into the thyrocyte is the first step in the formation of T3 and T4.
   - Thiocyanates and perchlorates block this step.
2. **Oxidation of iodide to inorganic iodine:** This step needs the enzyme peroxidase.
   - Drugs which block this stage (thioamides) are sulfonamide, PAS (para-amino-salicylic acid), carbimazole and propylthiouracil.
3. **Formation of iodotyrosines**
   - Iodine + Tyrosine = MIT (monoiodotyrosine) and diiodotyrosine (DIT)
   - This step is inhibited by thiourea group of drugs, i.e. carbimazole.
4. **Coupling reactions**
   - Coupling of two molecules of DIT results in T4 and one molecule of DIT and MIT results in T3.
   - This stage is blocked by carbimazole.
5. **Hydrolysis**
   - The hormones combine with globulin to form a colloid-thyroglobulin. They are stored in the thyroid gland and released as required by process of hydrolysis.
   - T3 is an important physiological hormone and is fast-acting (few hours). T4 is a slow-acting hormone and takes about 4–14 days to act.

THYROID FUNCTION TESTS

Serum T3 and T4 estimation is most commonly performed. Other tests are not commonly done and some of them are obsolete (Table 19.1).

1. **Serum T3:** Normal levels—1.5–3.5 nmol/L
   - Most (80%) T3 is produced by deiodination of T4 in the liver, muscle, kidney and anterior pituitary.
   - T3 is 3 to 4 times more potent than T4.
   - The half-life of T3 is approximately 24 hours, whereas half-life of T4 is about 7 days.

Table 19.1 Levels of T3, T4 and TSH in some common conditions

Disease	T3	T4	TSH
• Thyrotoxicosis	↑	↑	Suppressed or undetectable
• T3 toxicosis	↑↑	Normal	Suppressed
• Hypothyroidism	Low or normal	Low	↑

   - Free T3 (3 to 9 pmol/L) is most useful in confirming the diagnosis of early hyperthyroidism, especially in pregnancy where in levels of free T4 and free T3 rise before total T4 and T3.
2. **Serum T4:** Normal levels—55 to 150 nmol/L
   - They are measured by radioimmunoassay.
   - In euthyroid state, T4 is the predominant hormone produced by the thyroid.
   - Total T4 levels reflect output from the thyroid gland. Both T3 and T4 increase cell metabolism, normal growth, facilitate normal mental development and increase local effects of catecholamines.
3. **Serum TSH (thyroid stimulating hormone)**

   0.3–5 IU/ml of plasma. Table 19.1 shows the levels of T3, T4 and TSH in a few common conditions.
4. **Serum thyroglobulin**
   - It is produced by thyroid tissue only. Hence, the levels should be low after total thyroidectomy.
   - The most important use of this test is to monitor patients after total thyroidectomy for well-differentiated carcinoma.
   - It is not normally released into circulation in large amount but increases suddenly in thyroiditis, Graves' disease or toxic multinodular goitre (MNG).
5. **Serum cholesterol:** It is increased in hypothyroidism and decreased in hyperthyroidism.
6. **Thyroid autoantibody levels:** More than 90% of the patients with Hashimoto's thyroiditis and 80% of patients with Graves' disease have antibodies which are called as 'LATS' (long acting thyroid stimulator). The detection of these antibodies help in the diagnosis of such cases and also to suspect these diseases before clinical manifestation.
7. **Thyroid scintigraphy** (Table 19.2 and Fig. 19.7): Uptake by both lobes.

Table 19.3 shows a few terminology of the thyroid diseases.

Table 19.2 Thyroid scintigraphy

Substance	Dose	Half-life	Ideal case
^{131}I	High dose radiation (500 mrad)	Long 8–10 days	Lingual thyroid, retrosternal goitre; images are better
^{123}I	Low dose radiation (30 mrad)	Shorter 8–10 hours	Well differentiated carcinoma for bony metastasis.
^{99m} Tc (technetium)	Lowest radiation (5 mCi)	Shorter half-life 6–8 hours	Sensitive for nodal metastasis Lingual thyroid.

Fig. 19.7

Table 19.3 Nomenclature of certain thyroid diseases

• **Ectopic thyroid** Diagnosis by isotope scan or CT scan	• Thyroid tissue along the line of descent Example: Floor of the mouth, submental region, mediastinum
• **Lingual thyroid** Diagnosis by isotope scan or CT scan	• Swelling in the region of foramen caecum in tongue
• **Dyshormonogenesis** It is familial.	• Autosomal recessive condition with deficiency of peroxidase or dehalogenase (enzymes)
• **Pendred's syndrome**	• Dyshormonogenesis with congenital deafness
• **Struma ovarii**	• Malignant ovarian teratoma containing thyroid tissue
• **Jod Basedow's disease** (German word)	• Excessive iodine given for hyperplastic goitre resulting in hyperthyroidism

CLINICAL EXAMINATION OF THYROID SWELLING

Diseases of the thyroid are very common and thyroid swellings are very often common cases in an undergraduate and postgraduate clinical examination. Hence, before discussing the various diseases of the thyroid gland, various aspects of the "CLINICS" are discussed in detail below.

COMPLAINTS—HISTORY TAKING (Fig. 19.8)

1. **Swelling:** Long duration of thyroid swelling indicates benign condition, e.g. multinodular goitre (MNG), colloid goitre.
   - Short duration with rapid growth indicates malignancy such as anaplastic carcinoma. Majority of thyroid swellings do not produce pain.
2. **Rate of growth:** Usually slow-growing in benign disease.
   - If it is a rapid growth, it can be **'de novo' malignancy** or malignancy developing in a benign lesion, e.g. follicular carcinoma in MNG.
   - Sudden increase in the size of swelling with pain indicates haemorrhage in the MNG (multinodular goitre).
3. **Dyspnoea:** Difficulty in breathing in a patient with goitre can be due to following reasons (Key Box 19.2).
   - Small goitre, rapid growth—anaplastic carcinoma infiltrating the trachea.
   - When lower border is not seen, retrosternal goitre.
   - Hyperthyroidism causing arrhythmias leading to congestive cardiac failure can cause dyspnoea and orthopnoea.
   - Long-standing MNG compresses on the tracheal cartilages and produces **pressure atrophy** of tracheal cartilages. This is called **tracheomalacia**.
4. **Dysphagia** is relatively uncommon because oesophagus is a posterior structure and it is a fibromuscular tube.
5. **Hoarseness of voice** indicates malignancy. It always occurs in carcinoma thyroid infiltrating the recurrent laryngeal nerve (never in benign diseases of thyroid).
6. **Toxic features** suggestive of hyperthyroidism
   A. CNS symptoms are predominantly seen in Graves' disease (primary thyrotoxicosis—Key Box 19.3)
   - Tremors of the hand
   - Sweating
   - Intolerance to heat
   - Preference to cold
   - Excitability

KEY BOX 19.2

DYSPNOEA IN GOITRE—CAUSES

• Infiltration of trachea	Anaplastic carcinoma
• Lower border not seen	Retrosternal goitre
• Tracheomalacia	Long-standing MNG
• Cardiac failure	Secondary thyrotoxicosis

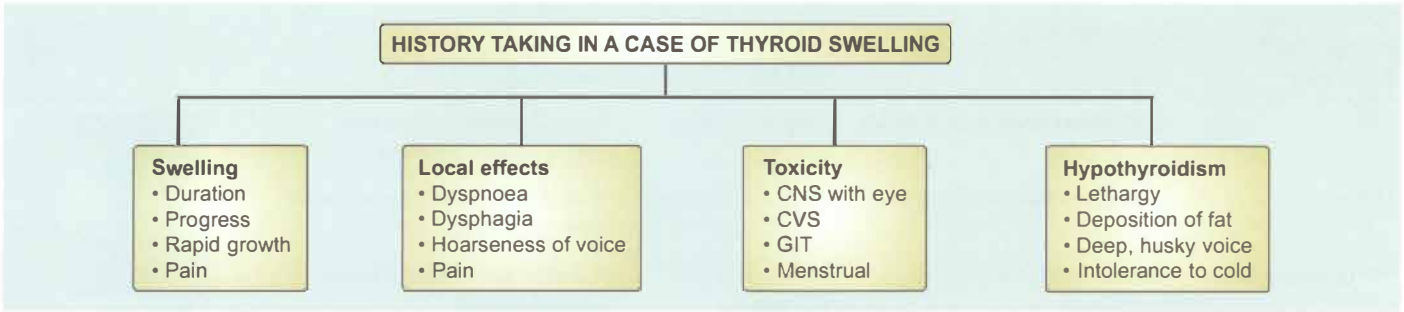


Fig. 19.8: Summary of history taking

KEY BOX 19.3

GRAVES' DISEASE

- Goitre
- Ophthalmic symptoms
- Irritability
- Tremors
- Restlessness
- Excitability

Remember as **GOITRE**

KEY BOX 19.4

SWELLINGS WHICH MOVE UPWARDS WITH DEGLUTITION

- Thyroid swellings
- Subhyoid bursitis
- Pretracheal and prelaryngeal lymph nodes
- Thyroglossal cyst
- Laryngocoele

- Irritability
- Prominent eyes are observed by other persons. Double vision, oedema of the conjunctiva can be the presenting complaints in late cases.

*B. **Cardiovascular symptoms*** (CVS) are predominantly seen in secondary thyrotoxicosis. Even though both forms of thyrotoxicosis produce palpitations, it is a significant complaint in multinodular goitre with thyrotoxicosis (secondary thyrotoxicosis). Precordial chest pain and dyspnoea on exertion are late manifestations of secondary thyrotoxicosis (Summary—Fig. 19.8)

ON EXAMINATION

Inspection

1. The location of the swelling is in front of the neck, extending from one sternomastoid to the other sternomastoid, vertically from suprasternal notch to the thyroid cartilage.
2. The size and shape have to be mentioned.
3. ***Surface:*** Thyroid swellings can have the following types of surfaces
   a. Smooth—adenoma, puberty goitre, Graves' disease
   b. Irregular—carcinoma of the thyroid
   c. Nodular—multinodular thyroid
4. Borders are usually round.
5. Swelling moves up with deglutition because of the following reasons (Key Box 19.4):
   - Thyroid is enclosed by pretracheal fascia which is condensed to form a ligament posteromedially called **ligament of Berry**. These are pairs of ligaments attached above to cricoid cartilage. During deglutition, the cricoid cartilage moves upwards and with it, the thyroid gland (give the patient a glass of water and check for movement with deglutition).
   - **If there is restriction of movement, it can be due to**
     - Malignancy with fixity to the trachea
     - Retrosternal goitre
     - Large goitre because of the size
     - Previous surgery
6. Movement on protrusion of the tongue suggests thyroglossal cyst. This test should be done when there is a nodule or a cyst in the region of isthmus of the thyroid gland. This test has no relevance in cases of MNG or other thyroid swellings.

Palpation

It should be done from behind.

1. Size, shape, surface and border should be confirmed. Local rise of temperature is a feature of toxic goitres. Very large nodular surface is described as bosselated surface (Figs 19.9 and 19.10).
2. Consistency
   - ***Soft:*** Graves' disease, colloid goitre.
   - ***Firm:*** Adenoma, multinodular goitre.
   - ***Hard:*** Carcinoma, calcification in the MNG.
3. Confirm the movement with deglutition by holding the thyroid gland.
4. **Intrinsic mobility** of the gland is very much restricted in carcinoma because of infiltration into the trachea.

Figs 19.9 and 19.10: Endemic goitre of 35 years duration turned into multinodular goitre. She presented with 3 months history of rapid increase in the size of the swelling. She underwent total thyroidectomy. Final histopathology report was follicular carcinoma thyroid. Observe large nodular surface—bosselations. (*Courtesy:* Dr Chidananda KV, Prof and Head of the Department and Dr Gopinath Pai, Prof of Surgery, KVG Medical College, Sullia, Dakshina Kannada, Karnataka. MBBS exam case 2007)

5. **Sternomastoid contraction test** is done when only one lobe is enlarged. In this situation the examiner keeps the hand on the side of the chin, opposite the side of the lesion and asks the patient to push his hand against resistance. If the gland becomes less prominent (as with thyroid swellings), it indicates the swelling is deep to the sternomastoid muscle.
6. **Chin test (neck fixation test)** is classically done in multinodular goitre, wherein both lobes are enlarged. The patient is asked to bend the chin downwards against resistance. This produces contraction of both sternomastoids and strap muscles and the gland becomes less prominent (Fig. 19.11).
7. **Special tests** or methods of examination of thyroid gland:
   a. **Crile's method** is indicated when there is a doubtful nodule. Keep the thumb over the suspected area of the nodule and ask the patient to swallow. The nodularity is appreciated better with this test (Fig. 19.12).
   b. **Lahey's method** of examination of thyroid can be done from front as well as behind. In order to palpate the right lobe, push the gland to the right side and feel the nodules in the posteromedial aspect of the gland. The lobe becomes more prominent and thus nodules are appreciated better.
   c. **Pizzillo's method is indicated in obese patients especially short-necked individuals.** The patient is asked to clasp her hands and press against her occiput with head extended. Thyroid gland becomes more prominent and thus, palpation becomes better.
   d. **Kocher's test:** If gentle compression on lateral lobes produces stridor, it is described as positive. This is due to **scabbard**[1] **trachea**. Long-standing multinodular goitres causing tracheomalacia and carcinoma with infiltration into trachea may give rise to stridor.
8. **Position of trachea:** In cases of solitary nodule confined to one lobe, trachea is deviated to the opposite side. However, in cases of multinodular goitres, trachea need not be deviated because of symmetrical enlargement of both lobes.
9. **Palpation of lymph nodes** in the neck. If lymph nodes are significant, it indicates papillary carcinoma of the thyroid.
10. **Palpation of common carotid artery:** Draw a line from mastoid process to sternoclavicular joint. Then draw a horizontal line from upper border of thyroid cartilage. The point where these two lines meet is the site of bifurcation of common carotid artery. This artery should be palpated just below this point.

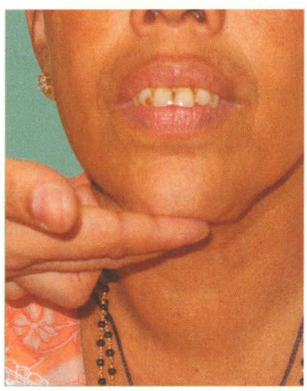


Fig. 19.11: Chin test—neck flexion test—thyroid swellings become less prominent

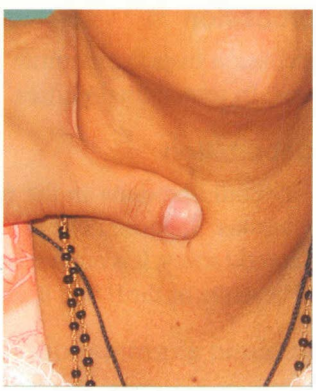


Fig. 19.12: Crile's method to detect nodule wherein thumb is used

[1]Scabbard is a sheath for holding a sword or other large blade (it is narrow and curved).

- In large multinodular goitres, the artery may be pushed laterally. Hence, pulsations are felt in the posterior triangle. Carcinoma of the thyroid engulfs the carotid sheath. Consequently, pulsations may be absent. Absent carotid artery pulsation is called **Berry sign positive**. Since the lumen is not narrowed, superficial temporal artery pulsations are felt normally.
- Summary of the palpation (Key Box 19.5).

KEY BOX 19.5

PALPATION TESTS

- Local rise of temperature, size, shape, surface, borders
- Consistency, movement with deglutition
- Intrinsic mobility test
- Sternomastoid contraction test and chin test
- Position of trachea
- Palpation of lymph nodes
- Pulsations of common carotid artery
- Special tests
- Evidence of toxicity

Percussion

Percussion over the sternum gives a resonant note in normal cases. In retrosternal goitres, it gives a dull note.

Auscultation

- It should be done in the upper pole because of following reasons: Superior thyroid artery is a direct branch of external carotid artery. It is more superficial than inferior thyroid artery.
- Presence of thrill and bruit are the features of toxic goitre.

Systemic examination

This includes CNS and eye signs, as in Graves' disease, examination of skeletal system to rule out metastasis as in carcinoma of the thyroid, and examination of cardiovascular system in cases of toxic goitre. These have been dealt with in detail in the corresponding topics. Deep tendon reflexes also have to be elicited—there is a slow relaxation phase in hypothyroidism.

Diagnosis

It is based on its anatomical location and features. It should be noted that the neural tumours arising from vagus nerve can present in the same location but it will not move with deglutition (Key Box 19.6).

KEY BOX 19.6

ANATOMICAL FEATURES OF THE THYROID GLAND

1. Thyroid gland is in front of the neck
2. Deep to pretracheal fascia
3. Moves up with deglutition
4. Butterfly shaped when whole gland is enlarged.

Differential diagnosis

- Simple goitre
- Toxic goitre
- Malignant goitre
- Solitary nodule
- Thyroiditis
- Other rare causes of thyroid enlargement.

GOITRE

Definition

Diffuse enlargement of the thyroid gland is described as goitre (it is derived from the Latin word, Guttur = the throat).

Classification of goitre

I. Simple goitre

- Puberty goitre
- Colloid goitre, iodine deficiency goitre (Figs 19.13 and 19.14)
- Multinodular goitre (Fig. 19.15).

II. Toxic goitre

- Graves' disease; diffuse toxic goitre
- Secondary thyrotoxicosis in multinodular goitre
- Toxic nodule; other causes.

III. Neoplastic goitre

- Benign adenoma (follicular adenoma)
- Malignant tumours: They are further classified as follows:

A. ***PRIMARY***

- Well-differentiated carcinoma
  - Papillary carcinoma, follicular carcinoma.
- Poorly differentiated carcinoma
  - Anaplastic carcinoma
- Arising from parafollicular cells
  - Medullary carcinoma
- Arising from lymphatic tissue
  - Non-Hodgkin's lymphoma.

B. ***SECONDARY (Metastasis)***

- Malignant melanoma, renal cell carcinoma, breast carcinoma produce secondaries in the thyroid, due to blood spread.

IV. Thyroiditis

- Granulomatous thyroiditis
- Autoimmune thyroiditis
- Riedel's thyroiditis.

V. Other rare causes of goitre

- Acute bacterial thyroiditis
- Thyroid cyst

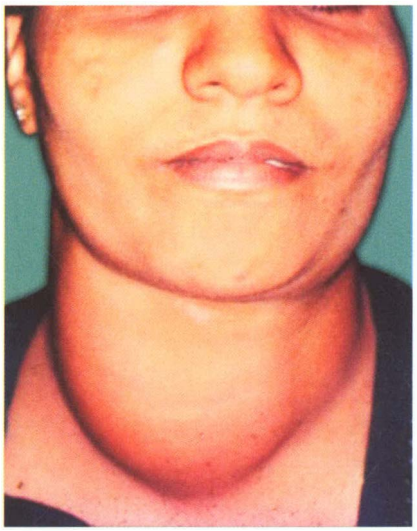


Fig. 19.13: Observe smooth surface—colloid goitre of 15 years duration

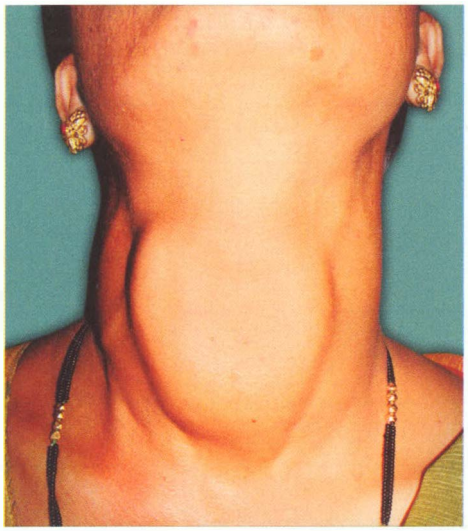


Fig. 19.14: Colloid goitre showing minor nodularity

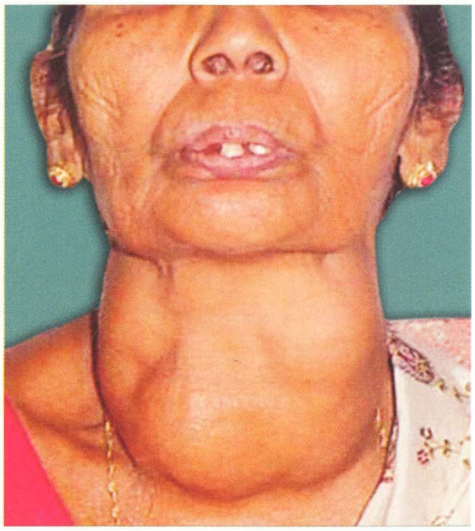


Fig. 19.15: Multinodular goitre showing nodules

- Thyroid abscess
- Amyloid goitres.

PEARLS OF WISDOM

Multinodular goitres, malignant goitres, puberty goitres are common causes of goitre. Bacterial thyroiditis is rare. Riedel's thyroiditis is very rare.

MULTINODULAR GOITRE

- A multinodular goitre is the end-stage result of diffuse hyperplastic goitre. Excessive metabolic demands cause an excessive enlargement of the thyroid. Therefore, it is common in women.
- Metabolic demands increase during puberty. A goitre appearing during that period is called puberty goitre. A goitre can develop during pregnancy and is called pregnancy goitre. Both of them are physiological but may eventually develop into multinodular goitre (MNG).

Aetiopathogenesis

Multinodular goitre occurs due to continuous stimulation by TSH secreted from the anterior pituitary.

1. Puberty goitre, pregnancy goitre

- It is seen in girls at puberty or during pregnancy when the metabolic demands are high and the production of T3, T4 are comparatively normal. Due to feedback mechanism, TSH levels increase, which stimulate thyroid gland and causes diffuse hypertrophy and hyperplasia.
- This is also called physiological goitre and can be treated by giving tablet thyroxine (T4) 0.2 mg/day to suppress TSH.
- Goitre may disappear if treatment is given in the stage of diffuse hypertrophy.

2. Iodine deficiency goitre

- Daily iodine requirement is about **100–125 micrograms.**
- Common in hilly/mountainous or low-lying areas because of decreased iodide content of water. This causes iodine deficiency goitre mediated by the same feedback mechanism.
- This is treated with iodised salt (which is used in food) and iodine—containing preparations.
- If the iodine deficiency status continues for a long time, it results in accumulation of colloid material in the gland and causes colloid goitre.
- All these three types of goitre if left untreated will change to multinodular goitre (Fig. 19.16).
  - **Stage I:** Stage of diffuse hypertrophy and hyperplasia of thyroid.
  - **Stage II:** Due to fluctuating levels of TSH because of pregnancy, lactation, menstruation, etc. Some areas in thyroid are overstimulated and are converted to active follicles.
  - **Stage III:** The active follicle ultimately undergoes necrosis and many such necrosed follicles join to form a nodule. Many such nodules form a multinodular goitre. Nodules contain necrosed tissue, i.e. inactive tissue. The internodular tissue is active.

3. Goitrogens such as cabbage (contains thiocyanates), drugs such as PAS and sulfonamides, cause goitre by preventing oxidation of iodide to iodine. Excess iodides inhibit organic binding of iodine and produce goitre.

4. Dyshormonogenesis (*see* Table 19.3)

Clinical features

- Common in females. Female : male ratio is 10 : 1. Seen in the age group of 20–40 years.

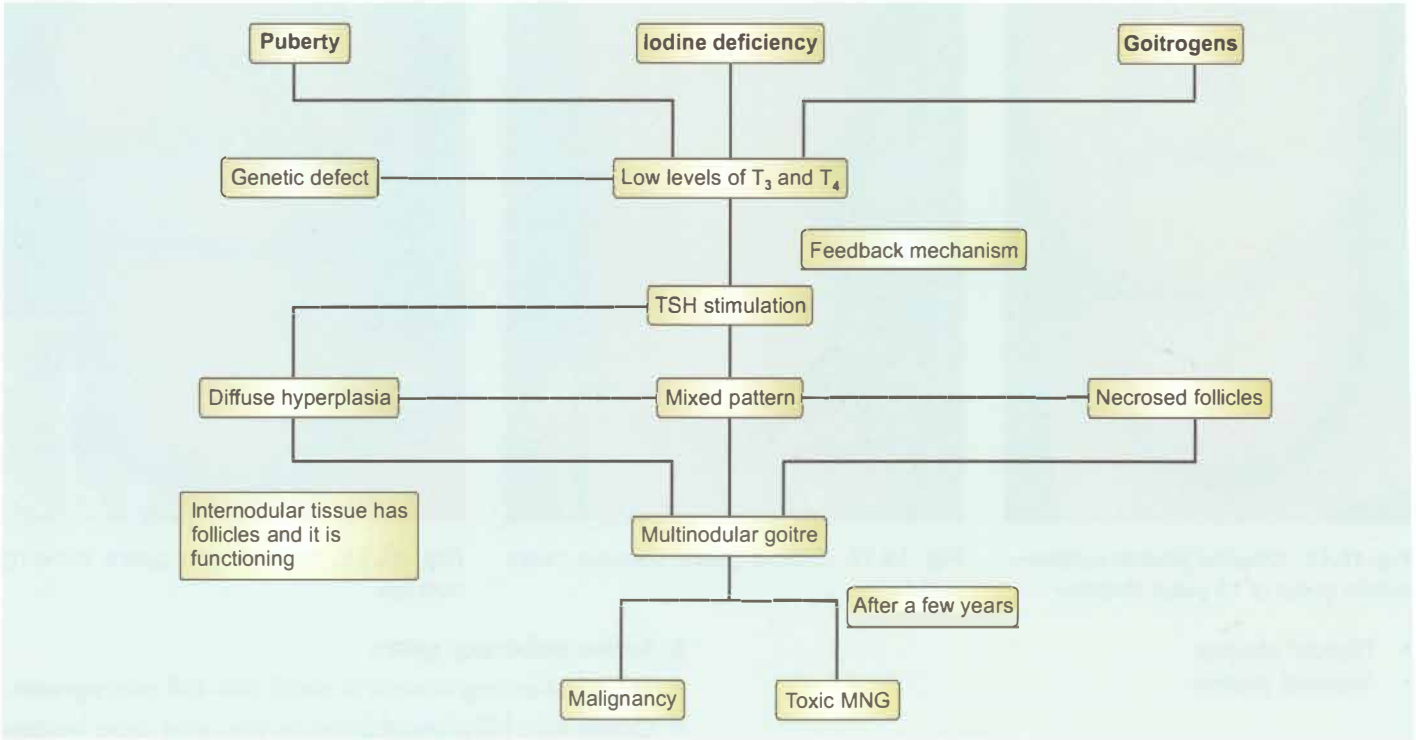


Fig. 19.16: Pathogenesis of MNG

- Long duration of swelling in front of the neck, dyspnoea due to tracheomalacia and dysphagia are the presenting features. The gland is nodular, firm in consistency and both the lobes are enlarged. Hard areas may suggest calcification and soft areas, necrosis.
- Sudden increase in size with pain is mainly due to haemorrhage in a nodule.

PEARLS OF WISDOM

The most common site of a nodule is at the junction of isthmus with one lobe.

Complications of multinodular goitre

1. Calcification in long-standing MNG.
2. Sudden haemorrhage in one of the nodules causes dyspnoea.
3. In 10–20% of cases, patients can develop secondary thyrotoxicosis with CVS involvement. Toxic multinodular goitre is also called **Plummer's disease**.

Tachycardia can be graded as follows—**Crile's** grading

Grade I	< 90/min—mild
Grade II	90 to 100/min—moderate
Grade III>	110/min—severe

4. Follicular carcinoma in a long-standing goitre (8%).

Management of multinodular goitre

Investigations

1. **Complete blood picture (CBP),** routine urine examination and fasting and postprandial blood sugar to rule out diabetes mellitus.
2. **X-ray of the neck:** Anteroposterior and lateral view.
   - To look for compression of trachea—to check feasibility of intubation during anaesthesia (Fig. 19.17).
   - To rule out retrosternal extension—soft tissue shadow seen.
   - Calcification in long-standing MNG.
3. **Flexible laryngoscopy** is done to see vocal cord mobility (this has replaced indirect laryngoscopy).
4. **Ultrasonography:** High frequency ultrasound is a very useful investigation specially in cases of solitary nodule. Even in multinodular goitres, ultrasound guided FNAC can be done. It can also detect clinically impalpable lymph nodes in the neck (suggest malignancy) (Key Box 19.7).
   - Ultrasound examination is inexpensive, easily done and has more advantages than disadvantages. Thus, it is often the first investigation in thyroid swellings.
5. **Fine needle aspiration cytology (FNAC):** It can be done in suspected hard nodule of multinodular goitre. It is a simple and useful investigation which can detect

Fig. 19.17: X-ray of the neck—lateral view

KEY BOX 19.7

ULTRASOUND

- It has become the investigation of choice (front line) in thyroid diseases.
- High frequency sonogram is used.
- It can distinguish solid from cystic lesions.
- It can measure the size of the nodule.
- It can reveal multicentric nature of the goitre.
- It can detect lymphadenopathy and can guide FNAC.
- It can detect microcalcification—a feature of malignancy
- ↑ echogenicity means ↑ chances of tumour/malignancy
- ↑ sonolucent means ↑ chances of cyst (benign).

malignancy. Since the treatment of MNG is often total thyroidectomy and an ultrasonogram can also help to rule out malignancy, ***FNAC is done only in suspicious cases of MNG.***

6. CT scan is done when you suspect retrosternal extension, doubtful resectability or large lymph nodes in the neck. They may also have intrathoracic lymph nodes (Fig. 19.55).

Classification of investigations (Key Box 19.8)

Prevention

Prevention is mainly important in endemic area and is done by supplementing iodine (Key Box 19.9).

KEY BOX 19.8

INVESTIGATIONS—GOITRE

1. Simple goitres
   - Routine—blood and urine tests, chest X-ray
   - Indirect laryngoscopy—flexible laryngoscopy is better
2. Toxic goitres
   - Routine
   - T3, T4, TSH
3. Malignant goitre
   - Routine
   - Ultrasound, CT scan of the neck
   - FNAC

KEY BOX 19.9

PREVENTION OF MNG

- ***Puberty goitre:*** 0.1 mg to 0.2 mg of thyroxine
- ***Iodine deficiency goitre:*** Use iodised salt, sea food, milk, egg, etc.
- ***Goitrogens:*** Avoid cabbage, drugs.

Why do we ask for ultrasound examination?

Following clinical notes will give answers.

CLINICAL NOTES

1. *A 22-year-old lady was referred to us for a nodule in the right side of neck in the thyroid region that was moving with deglutition. She also had epigastric pain since 6 months. Clinical diagnosis of solitary thyroid nodule with hyperacidity was made. An ultrasound examination of the neck was requested and the report was a surprise. She was having a parathyroid adenoma with calcific pancreatitis and nephrocalcinosis. You know the diagnosis now—A case of hyperparathyroidism.*
2. *We sent a case of MNG to thyroid scan. The scan detected jugular lymphadenopathy on both sides. The diagnosis changed from MNG (benign) to malignancy (papillary carcinoma because of lymph nodes). Lymph nodes were not palpable on clinical examination in this patient.*
3. *A 21-year-old lady had a multinodular goitre with the entire gland replaced by nodules. Case was posted for a total/near total thyroidectomy. A senior experienced Professor asked one question, 'what is the duration of this swelling?' It was 10 years. As per his advice, FNAC was done. FNAC report was lymphocytic thyroiditis. Surgery was cancelled. She was put on T-Eltroxin 0.1 mg, 2 tablets /day. She is on follow up now. Gland size is reduced to 30% in the last 2 years.*

Treatment

Three choices can be given to patients.

- **Subtotal thyroidectomy:** In this operation, parts of right and left lobes and entire isthmus are removed in flush with tracheal surface leaving behind a little tissue in the tracheoesophageal groove to protect recurrent laryngeal nerve and parathyroid gland (Fig. 19.18).
- Some surgeons treat these patients with 0.2 mg of thyroxine to suppress the TSH stimulation in the postoperative period, for a period of 2–5 years.
- **Total thyroidectomy is the choice today provided** complications such as recurrent laryngeal nerve, paralysis and hypocalcaemia due to removal of parathyroid glands can be avoided. Thus, it is desirable to do a total thyroidectomy if **experience of the surgeon is good** and in a **high volume centre** (Fig. 19.19).
- **Medical:** Small nodules—treat with Tab thyroxine (Eltroxin) 0.1 to 0.2 mg/day.

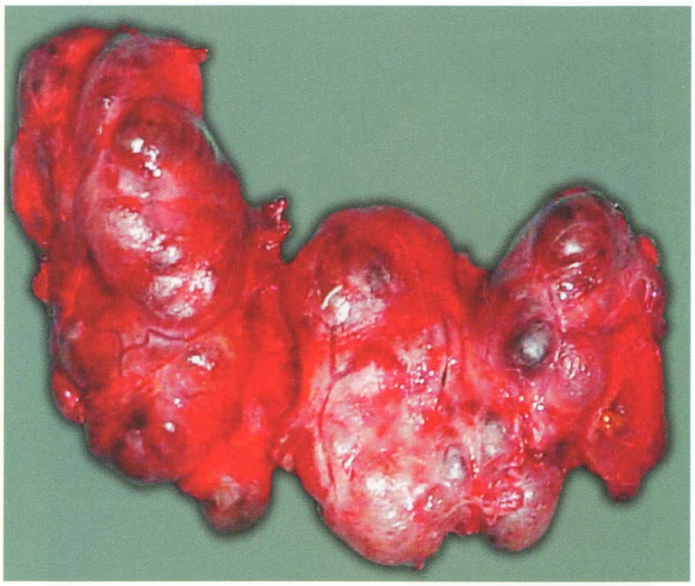


Fig. 19.18: Subtotal thyroidectomy specimen

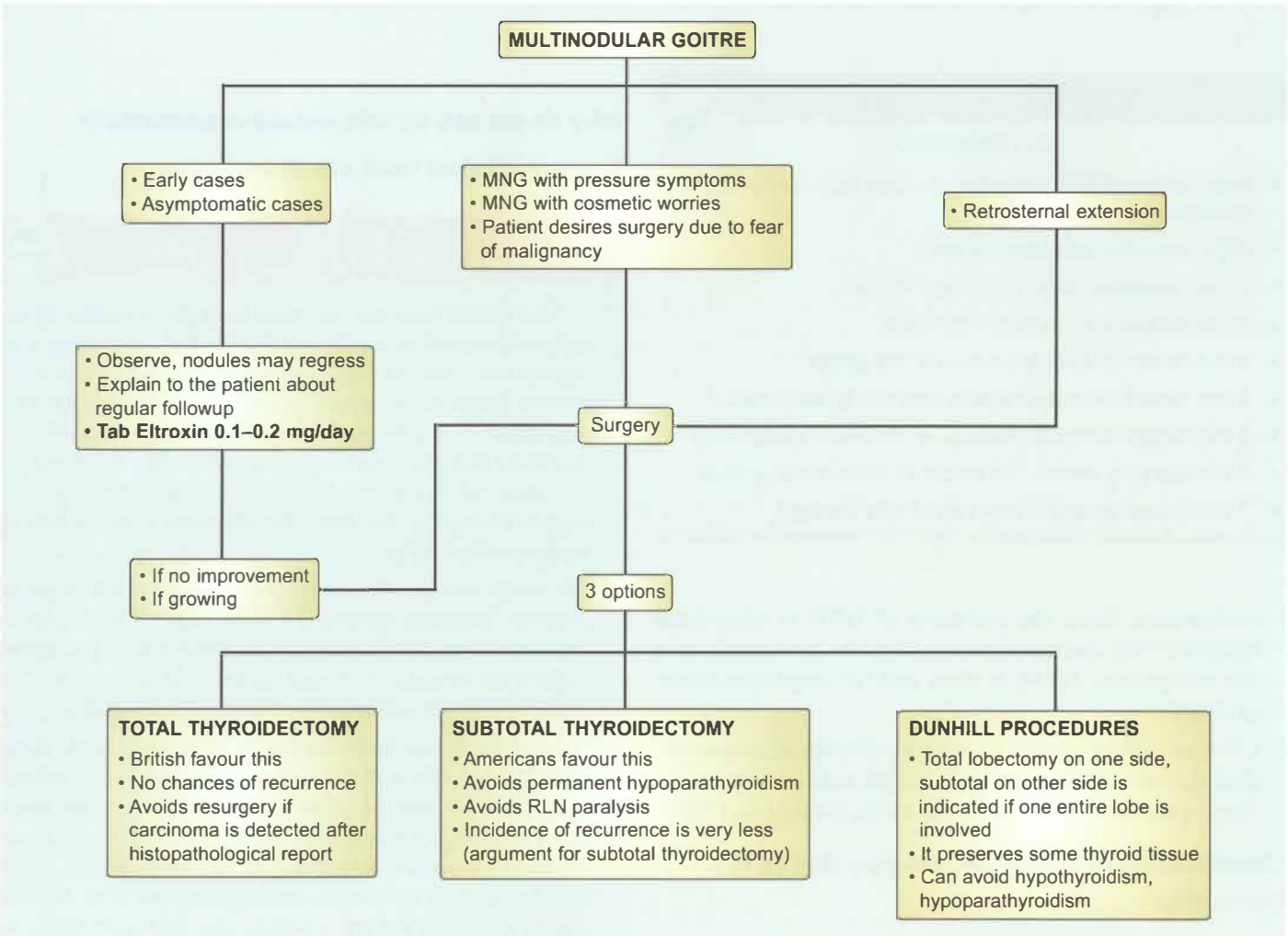


Fig. 19.19: Management of multinodular goitre—for diagrams *see* operative surgery chapter on thyroid diseases

RETROSTERNAL GOITRE

Very often, it is a multinodular goitre that develops in the neck and is slowly pulled down behind the sternum due to following reasons.

1. Negative intrathoracic pressure
2. Pretracheal muscles are strong in men
3. Short neck, obesity

Rarely, it arises from an ectopic thyroid tissue.

Classification

I. **Primary:** It arises from ectopic thyroid tissue in the mediastinum. It also derives blood supply from mediastinum. It is rare (1%)

II. **Secondary:** It is the common variety—It is MNG which gets pulled down into thorax.

Clinical types

- **Substernal:** The most common type where the lower border of the gland is behind the sternum.
- **Intrathoracic:** No thyroid is seen in the neck, diagnosed by radio-iodine scan.
- **Plunging goitre:** When patient is asked to cough, intrathoracic pressure increases. As the thyroid plunges out, the lower border of gland is clearly seen in the neck.

Clinical features of retrosternal goitre

- It can be suspected when the lower border of the swelling is not seen.
- Most of the patients have difficulty in breathing or even stridor.
- Dysphagia is more common.
- Engorgement of neck veins and superficial veins. These become more prominent when the hands are raised above the head, and the arms touch the ears—**Pemberton's sign** (Fig. 19.20).

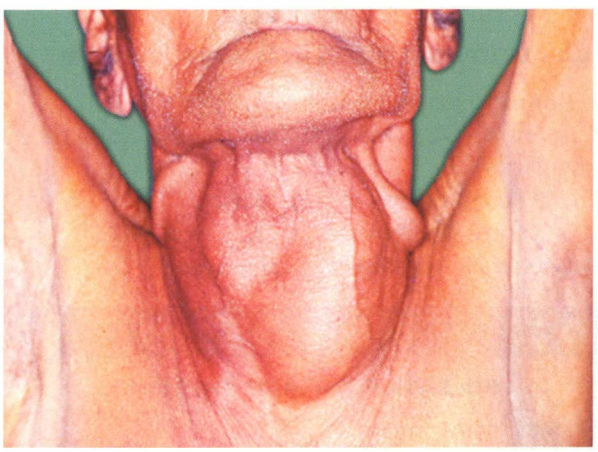


Fig. 19.20: Observe the engorged veins due to retrosternal goitre. Such veins are also seen when mediastinal nodes are enlarged as in papillary carcinoma thyroid

Investigations

- They are similar to MNG. However, isotope scan is very useful in the diagnosis of intrathoracic goitres.
- CT scan to localise and to know the size and extent

Treatment

- It can be easily explored through the neck incision and removed.
- Very rarely, a sternal split may be necessary.
- **Summary** (Key Box 19.10).

KEY BOX 19.10

RETROSTERNAL GOITRE

- Very often, it is an MNG with the lower border unseen
- Rarely from ectopic thyroid tissue
- Severe breathlessness even though small
- Drugs should not be given if it is toxic
- Pressure effects diagnosed by **Pemberton's test**
- Surgical excision is the treatment
- No radioiodine therapy

TOXIC GOITRE—THYROTOXICOSIS

It is a complex disorder which occurs due to increased levels of thyroid hormones (hyperthyroidism) and manifests clinically with various signs and symptoms involving many body systems. Following are the causes of thyrotoxicosis:

1. **Primary thyrotoxicosis** (Graves' disease, exophthalmic goitre, diffuse goitre)
2. **Secondary thyrotoxicosis:** Secondary to nodular goitre (multinodular)
3. **Solitary toxic nodule:** Autonomous nodule which is not under the influence of TSH but occurs due to hypertrophy and hyperplasia of gland (tertiary thyrotoxicosis).
4. **Other causes of thyrotoxicosis**
   - **Thyrotoxicosis factitia:** False thyrotoxicosis occurs due to overdosage of thyroxine, given for puberty goitre.
   - **Jod-Basedow's thyrotoxicosis:** Jod means iodine in the German language, Basedow means toxic goitre. Iodine-induced thyrotoxicosis (iodine given for hyperplastic endemic goitres).
   - Initial stage of **thyroiditis:** Hashimoto's thyroiditis, viral thyroiditis can produce temporary thyrotoxicosis features.
   - Very rarely, malignant goitres can be toxic (differentiated carcinoma).
   - **Neonatal thyrotoxicosis** occurs in babies born to thyrotoxic mothers.
   - TSH-secreting **tumours of pituitary**
   - **Struma ovarii**

- ***Drugs:*** Amiodarone is an iodine containing preparation given as antiarrhythmic drug.

Please note: In the clinical examination, primary thyrotoxicosis and secondary thyrotoxicosis are commonly kept as long cases. Hence, these two have been discussed below in detail and also *see* Table 19.5.

GRAVES' DISEASE

Aetiopathogenesis

The exact aetiological factors responsible for the disease are not clear. Following are considered as possible aetiological factors (Key Boxes 19.11 and 19.12):

1. **Autoimmune disorder** is the first possible cause due to the demonstration of auto-antibodies in the circulation. *Example:* TSH receptor antibodies. It can also be associated with other autoimmune disorders like vitiligo.
2. **Familial:** The disease can run in families. Familial/genetic Graves' disease has been documented in identical twins.
3. **Thyroid stimulating immunoglobulins** (TSI) and long-acting thyroid stimulator (LATS) are responsible for pathological changes in the thyroid gland in Graves' disease. They stimulate thyrocytes to grow and synthesise excess thyroid hormones.
4. **Exophthalmos producing substance (EPS)** is responsible for "ophthalmopathy" seen in Graves' disease.
5. **Female sex**, emotions, stress, young age also have been considered as other factors responsible for the disease.

Pathology

- As a result of continuous stimulation, acinar hypertrophy and hyperplasia take place. The acinar cells which are normally flat, become tall columnar. The normal colloid disappears and the cells are empty. However, rich vascularity is seen. Thus, small follicles with hyperplastic columnar epithelium is characteristic.

KEY BOX 19.11

TRIGGERING FACTORS FOR GRAVES' DISEASE

- Postpartum state
- Iodine excess
- Lithium therapy
- Infection—bacterial and viral

KEY BOX 19.12

FEATURES OF GRAVES' DISEASE

- **F**emale with strong family predisposition
- **E**xtrathyroidal manifestations
- **M**iddle or young age (30 to 50 years)
- **A**utoimmune mechanism
- **L**eukocyte antigen human (HLA) and T-lymphocyte antigen may contribute.
- **E**nlargement of gland is diffuse

Remember as **FEMALE**

CLINICAL NOTES

Following are three case reports which highlight the clinical symptomatology of primary thyrotoxicosis.

1. *An 18-year-old girl visited many doctors for her complaint of loss of weight. She was investigated for tuberculosis (common disease in India), malignancy, etc. She was given unnecessary tonics. After nearly 6 months, when eye-signs started developing, it was proved to be Graves' disease.*
2. *A bank clerk's only complaint was that he could not sign the cheque because of excessive sweating. Thyroid gland was not palpable. His pulse rate was very high, investigations revealed that it was a case of primary thyrotoxicosis. On careful questioning, he admitted that he was a "nervous character".*
3. *A 24-year-old lady was being asked by her friends every day why her eyes were prominent. Her only complaint was prominent eyes. On careful questioning, she admitted having anxiety, tension, excitability.*

Clinical features (*see* Clinical Notes)

- Primary thyrotoxicosis is 8 times more common in females than in males, especially in the age group of 15–25 years.
- **Symptoms, signs and swelling appear simultaneously.**
- Very often young women present with unexplained loss of weight in spite of good appetite. Diarrhoea occurs due to increased smooth muscle activity of small intestines. Intolerance to heat, preference to cold, fine tremors, excitability, hyperkinetic movements, excessive sweating are the other features. **Free steroid hormone levels decrease Graves' disease. This results in decreased effective oestrogen at the cell level which in turn causes oligomenorrhoea.**

Signs of primary thyrotoxicosis

I. Signs of thyroid gland in Graves' disease

- Uniformly enlarged (mild degree)
- Smooth surface—no nodules (treated cases may have nodularity)
- Gland is soft or firm in consistency
- It is warm—highly vascular (Key Box 19.13)
- Auscultation—a bruit is usually heard.

KEY BOX 19.13

PULSATILE THYROID SWELLINGS

- Primary thyrotoxicosis
- Secondary thyrotoxicosis
- Follicular carcinoma
- Vascular malformations

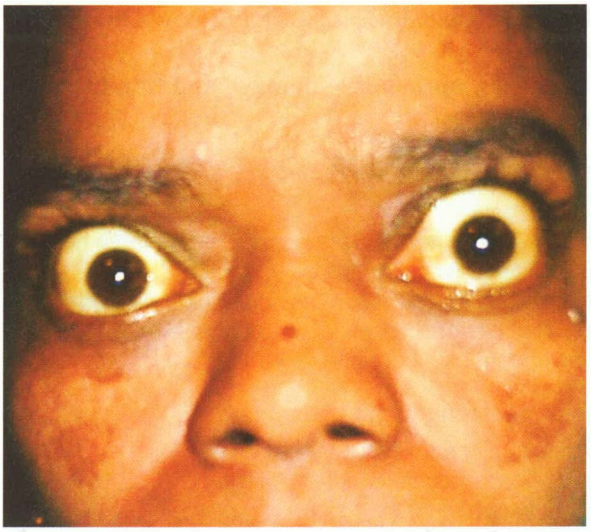


Fig. 19.21: She was 42-year-old with a 2-month history of thyrotoxicosis that developed after viral infection. (*Courtesy:* Dr Sreejayan, Prof. of Surgery, Calicut Medical College, Calicut, Kerala)

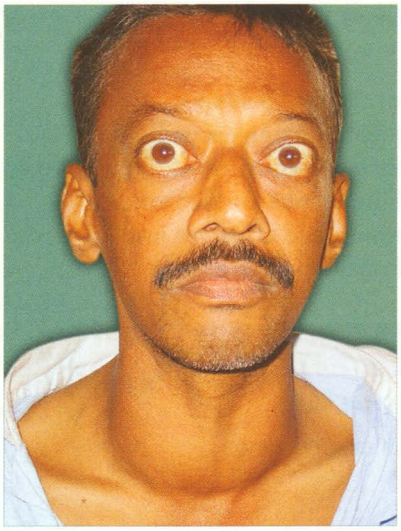


Fig. 19.22: He was 31-year-old with loss of weight of 12 kg in 3 months. There was diffuse enlargement of the gland with exophthalmos

Fig. 19.23: She was 48-year-old lady who had multinodular goitre of 10 years duration. She also had lid-lag sign (rare in toxic MNG)

II. Central nervous system (CNS) signs

- Tremors of the tongue when the tongue is within the oral cavity and tremors of the outstretched hands are characteristics. A piece of paper may be placed on the fingers in doubtful cases for demonstrating the tremors of the hand. Extensor surface of the hand is used because extensors are weak when compared to flexors.
- Hyperkinetic movements.
- Always a moist, warm hand (shake hands).

III. Cardiovascular system (CVS) signs

- Pulse rate is always raised and rapid indicating tachycardia. Depending upon the pulse rate, thyrotoxicosis can be classified as follows: mild—90–100/minute, moderate—100–110/minute, severe—more than 110/minute.
- Palpitation and extrasystoles can also be found in primary thyrotoxicosis even though other cardiac features such as fibrillation and cardiac failure are rare.

IV. Eye signs

- Prominent eyeballs and retraction of the eyelid result in thyrotoxic exophthalmos. This is due to retrobulbar deposition of inflammatory cells and round cells with venous congestion resulting in oedema (Figs 19.21 to 19.23).
- **Levator palpebrae superioris** muscle is innervated by **oculomotor nerve** which also carries sympathetic fibres derived from cavernous plexus for the smooth muscle part of the levator. Contraction of this muscle produces lid spasm.
- This is aided by **spasm of Müller's muscle**, a sympathetic muscle which lies adjoining the levator palpebrae superior muscle. This is responsible for keeping the eyeball forwards. All these factors together produce a classical stare.

1. **Assessment of exophthalmos**
   - Upper sclera is seen above the limbus (upper margin of the cornea and conjunctiva—**Dalrymple's sign**.
   - **Naffziger's method:** Stand behind the patient and look at the supraciliary arch, by tilting the patient's head backwards. In normal cases, eyeball is not seen. In cases of exophthalmos, eyeball is protruded outside and hence it is seen.
2. **Moebius' sign:** Loss of convergence of eyeball occurs due to muscle paresis as a part of thyrotoxic ophthalmoplegia. Diplopia is due to weakness of extraocular muscles (inferior oblique-elevators).
3. **Stellwag's sign:** Infrequent blinking and widening of palpebral fissure is due to spasm of sympathetic fibres in the levator palpebrae superioris.
4. **Joffroy's sign:** Absence of wrinkling of the forehead when the patient is asked to look upwards. This occurs due to increase in the field of vision due to exophthalmos.
5. **von Graefe's sign (Lid lag sign):** When the patient is asked to look up and down, upper eyelid cannot cope up with the speed of movement of the finger because of the lid spasm. Hence, the lid lags behind.
6. **Enroth sign:** Oedema of eyelids and conjunctiva
7. **Gifflord's sign:** Difficulty in everting the upper eyelid.
8. **Kocher's sign:** When an attempt is made to lift the eyes higher, upper eyelid springs up more quickly than the eyebrows

Summary (Table 19.4 and Key Boxes 19.14 to 19.16)

Malignant exophthalmos

- This occurs in untreated cases of Graves' disease.
- If the disease continues, infrequent blinking secondary to exophthalmos results in constant **exposure of the cornea to the atmosphere**. This results in keratitis, corneal ulcer, conjunctivitis, chemosis and may even lead to blindness. This is called malignant exophthalmos.

KEY BOX 19.14

THYROTOXIC EXOPHTHALMOS

- Proptosis and lid retraction result in exophthalmos
- Sclera is visible beyond limbus
- Naffziger's method to examine
- Staring look
- Typically seen in Graves' disease
- Rarely seen in secondary thyrotoxicosis

KEY BOX 19.15

INTERESTING 6 Ps OF GRAVES' OPHTHALMOPATHY

- **P**rominent eyes
- **P**eriorbital oedema
- **P**apilloedema
- **P**roptosis
- **P**alpebral fissure widening
- **P**rogression to blindness

KEY BOX 19.16

GRADING OF THYROID EYE DISEASES

Grade 0	No signs or symptoms
Grade 1	Only signs, no symptoms
Grade 2	Both signs and symptoms
Grade 3	Proptosis
Grade 4	Extraocular muscle involvement
Grade 5	Corneal involvement
Grade 6	Loss of vision with optic nerve atrophy

- Malignant exophthalmos is probably due to **autoimmune disease**.
- In late stages, **optic nerve damage and blindness** can occur.

Important causes of exophthalmos

1. Thyrotoxicosis
2. **Primary CNS tumours:** Meningioma, optic nerve glioma, haemangioma, lymphoma, etc.
3. **Metastatic tumours:** Neuroblastoma, central carcinoma.
4. **Vascular:** Cavernous sinus thrombosis—aneurysm of ophthalmic artery

Causes of pulsating exophthalmos

1. Cavernous sinus thrombosis
2. Carotid—cavernous sinus, A-V fistula
3. Orbital vascular tumour
4. Ophthalmic artery aneurysm

Treatment of thyrotoxic ophthalmopathy

1. Massive doses of steroids—methylprednisolone and metronidazole
2. Lateral tarsorrhaphy
3. Orbital decompression may be necessary in late cases
4. Guanethidine eyedrops are useful to decrease lid spasm and lid retraction.
5. Head end elevation and disease control.
6. Dark spectacles, 7% methylcellulose eye drops.

V. Thyrotoxic myopathy

- **Mild weakness of proximal limb muscles,** ocular and frontalis muscles is not uncommon. On careful questioning, patient may admit difficulty in climbing steps.
- Weakness of extraocular muscles results in double vision (diplopia).
- Features suggestive of myaesthenia gravis, periodic paralysis can be found.
- **Myopathy improves with treatment.**

VI. Thyrotoxic dermopathy (Key Box 19.17)

- Popularly called as pretibial myxoedema—is seen in thyrotoxicosis patients treated with surgery or antithyroid

Table 19.4 Symptoms of thyrotoxicosis

Symptoms of hyperthyroidism	Symptoms of increased adrenergic stimulation	In females	In children
Heat intolerance	Palpitations	Amenorrhoea	Rapid growth
Increased sweating, thirst	Nervousness, fatigue	Decreased fertility	
Weight loss in spite of good caloric intake	Emotional lability	Miscarriages	
	Hyperkinesis		
	Tremors	**Prominent stare is due to catecholamine excess.**	

drugs. It is always associated with exophthalmos. It is seen in 1–2% of patients.

- Bilateral symmetrical deposition of myxomatous tissue (glycosaminoglycans) mainly in the pretibial region, may also affect the foot and ankle, sometimes the entire leg below knee. Skin is dry and coarse (thickening of skin by mucin—like deposits). **Swelling is due to the obliteration of initial lymphatics by mucin** (*see* page 111).
- Pretibial myxoedema (misnomer) is nonpitting in nature and may be associated with clubbing of fingers and toes called thyroid acropachy (Key Boxes 19.18 and 19.19).
- Responds to topical steroids and thyroid disorder treatment

Thus 4 important features are seen only in primary thyrotoxicosis, not in secondary thyrotoxicosis (Key Box 19.20).

KEY BOX 19.17

SKIN CHANGES

- Pretibial myxoedema
- Pruritis
- Palmar erythema
- Thinning of hair
- Dupuytren's contracture

KEY BOX 19.18

PRETIBIAL MYXOEDEMA—MISNOMER

- **A**cropachy—Clubbing of fingers and toes
- **C**oarse hair
- **R**ed shiny skin
- **O**bliteration of initial lymphatics by mucin, oedema non-pitting
- **P**retibial region, foot and ankle
- **A**fter a few years of toxicosis, it develops
- **C**yanotic when cold
- **H** } aluronic acid deposition
- **Y** } in dermis—bilateral and symmetrical

Remember as **ACROPACHY**

KEY BOX 19.19

SOME MISNOMERS

• Pretibial myxoedema	–	Not seen in myxoedema
• Mycosis fungoides	–	Not a fungal infection
• White bile	–	Not white, not bile
• Adenolymphoma	–	Not a lymphoma
• Sternomastoid tumour	–	Not a tumour
• Malignant hydatid	–	Not malignant

KEY BOX 19.20

EXTRATHYROIDAL MANIFESTATIONS OF GRAVES' DISEASE

- Pretibial myxoedema
- Proximal myopathy
- Pachy (acropachy)
- Progressive ophthalmoplegia

At the end of clinical examination, commonly asked question is, how will you differentiate primary thyrotoxicosis from secondary thyrotoxicosis (Table 19.5).

Management of primary thyrotoxicosis

Routine isotope scanning has been abandoned in toxic goitres except when toxicity is associated with nodularity.

Investigations

- Routine investigations such as complete blood picture, fasting and post-prandial blood sugar estimation, **flexible laryngoscopy** are done.
- Serum T3, T4 and TSH are measured. T3 or T4 levels are high and TSH levels are low. The normal level of T3 is 1.3–3.5 nmol/L and normal level of T4 is 55–150 nmol/L.
- **Sleeping pulse rate** is counted after the patient is sedated with 30 mg of phenobarbitone. In a case of toxic goitre, the pulse rate remains high even during sleep because of increased metabolism. This is a simple bedside

Table 19.5 Differences between primary and secondary thyrotoxicosis

	Primary thyrotoxicosis (Graves)	Secondary thyrotoxicosis (Toxic MNG)
1. Age	20–40 years	35–50 years
2. Symptoms and signs	Appear simultaneously, duration is short	Long duration of a swelling and short duration of signs
3. Skin over thyroid	Warm	Not warm
4. Consistency	Soft or firm	Firm or hard
5. Surface	Smooth	Nodular
6. Auscultation	Bruit is common	Bruit uncommon
7. Eye signs	Commonly found	Rarely found (lid lag)
8. Predominant symptoms	CNS	CVS
9. Pretibial myxoedema	Seen in 1–2% patients	Never seen
10. Proximal myopathy	Seen in 5% patients	Never seen
11. Malignant exophthalmos	Can be seen	Never seen

investigation in cases of toxic goitre. In anxiety states, pulse rate may be high in the waking hours and it comes back to normal during sleep.

- Thyroid stimulating antibodies are elevated (TSH-RAbs) Measurement of IgG immunoglobulins (TSH-RAbs) is not essential to make the diagnosis of thyrotoxicosis.

Treatment of primary thyrotoxicosis

Aim of treatment

I. To restore patients to euthyroid state
II. To reduce the functioning thyroid mass to a very critical level (about 6–8 g of thyroid tissue)
III. To minimise complications

I. To restore the patient to euthyroid state (Table 19.6)

- Other drugs such as **potassium perchlorate** are given in the dose of 200 to 400 mg daily. Propylthiouracil in the dose of 200 mg three times a day can also be given in patients **who develop neutropaenia due to carbimazole**.
- Propranolol inhibits peripheral conversion of T4 to T3. This results in rapid control of tachycardia and surgery can be scheduled in a few days (within one week). It should be continued for one week after surgery because it does not interfere with synthesis of hormones.

Please note

- Iodine containing **antiarrhythmic drug amiodarone** may worsen thyrotoxicosis.
- **Propyl thiouracil is safe in pregnancy** with Graves' disease.
- **Role of Lugol's iodine is doubtful.**
- **Antithyroid drugs will not cure** the disease. In selected patients (30–40%) remission is possible with regular intake of drugs. They may be continued for a maximum period of 2 years. If toxicity persists or recurs on stopping drugs, surgery is recommended. However, majority of the patients ultimately require surgery or radioiodine.

Block and replace treatment

- If a small dose of T3 (20 μg up to 4 times/day) or T4 (0.1 mg/day) is given along with antithyroid drugs, there is less incidence of development of hypothyroidism and increase in the size of goitre.

II. To reduce the functioning thyroid mass

1. ***Subtotal thyroidectomy:*** In this procedure, the amount of thyroid gland removed is more than toxic multinodular goitre. However, being a toxic goitre, very little gland should be left behind so as to avoid persistent toxicity in the postoperative period. This quantity is difficult to measure. Hence, some surgeons advocate leaving thyroid tissue as small as the tip of the little finger, on both sides. By convention it is said that a total of about 6–8 g of the thyroid tissue should be left on both sides of the tracheo-esophageal groove.
2. ***Total thyroidectomy*** can be offered to young patients with a small-sized gland. It controls the toxicity very faster. Hypothyroidism occurs but it is easy to treat.
3. ***Radioiodine therapy:*** This is a suitable alternative to surgery in cases of primary thyrotoxicosis in patients above the age of 30 (Key Box 19.21).

III. To minimise complications

Good preoperative preparation of the patient, good anaesthetic and surgical techniques, and good postoperative care will reduce the complications of surgery.

Thus, antithyroid drugs, subtotal thyroidectomy and radio-iodine therapy are the three different modalities available for the treatment of primary thyrotoxicosis. The indications, merits and demerits of each treatment are given in Fig. 19.24.

Treatment of secondary thyrotoxicosis

- Patients with severe cardiac damage entirely or partly due to hyperthyroidism are middle-aged or elderly with secondary thyrotoxicosis and the hyperthyroidism is not very severe. These patients develop atrial fibrillation and cardiac failure, if left untreated. In elderly patients, when the operative risk is unacceptable, radioiodine is given. Treatment with antithyroid drugs is started 48 hours later and continued until radioiodine has had its effect (6 weeks).
- If the cardiac symptoms are controlled well and anaesthesia risk is acceptable, subtotal thyroidectomy is done. However, the gland that is left behind should be equal to the distal phalanx of the thumb of the patient.

Table 19.6 Antithyroid drugs: Routine preoperative preparation

Drugs and mode of action	Dose	Precautions/Side effects
• Carbimazole: It blocks oxidation of iodide to iodine and coupling reactions thus reducing T3 and T4 levels. It is metabolised to methimazole after ingestion	10 mg, 6th hourly and maintenance dose of 10 mg two to three times a day	Takes 12 weeks for its action It should be given 3–8th hourly interval Dangerous agranulocytosis can manifest as sore throat
• Propranolol is a nonselective β-blocker, reduces tachycardia.	20–40 mg, two or three times a day depending on the pulse rate	Congestive cardiac failure Can precipitate bronchial asthma. T3 and T4 levels are not decreased
• Lugol's iodine is given to reduce vascularity of the gland before surgery	10–12 drops (minimum three times a day for 10 days before surgery)	It is bitter, has to be taken with orange juice

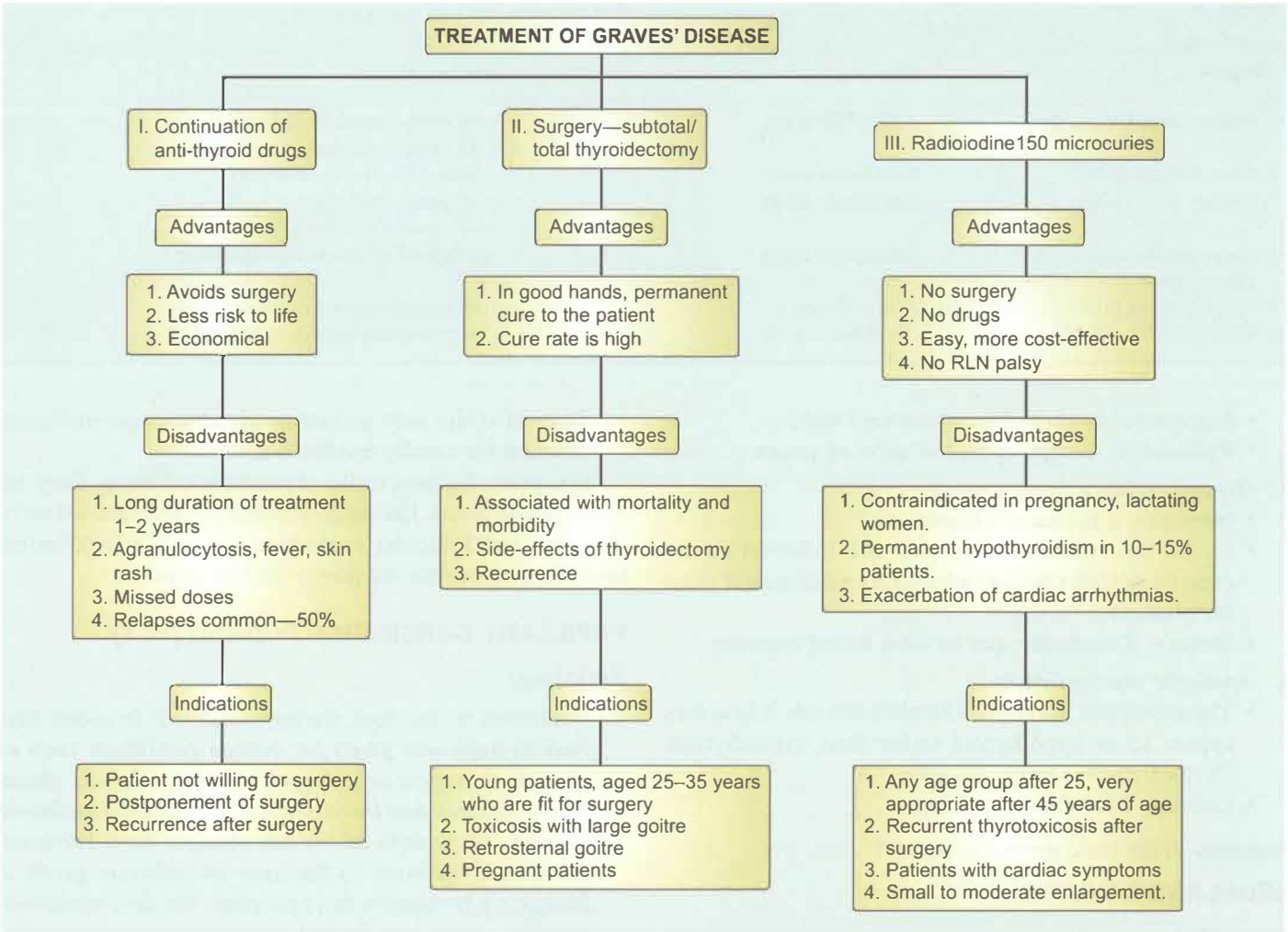


Fig. 19.24: Indications, merits and demerits of different modalities of treatments for primary thyrotoxicosis

Solitary toxic nodule

Treatment

- This is not under control of TSH. It is an automous nodule
- The control of toxicity is in the usual manner—carbimazole and propranolol
- This is an indication to do I^{131} scan or preferably IV 99mTc scan. Hot nodule takes up isotope (also in carcinoma). Rest of the gland does not.
- Treatment is either by hemithyroidectomy or radioactive iodine therapy—I^{131}.

Some specific thyroid conditions

1. ***Thyrotoxicosis in children:*** Initially antithyroid drugs are given for 10–15 years followed by surgery.
   - Radioiodine is absolutely contraindicated as there is a fear of carcinoma and growth retardation (Key Box 19.21).
2. ***Thyrocardiac:*** It refers to a condition wherein cardiac damage has resulted due to hyperthyroidism (Key Box 19.22).
   - Classically it happens in secondary thyrotoxicosis. It is usually seen in middle aged or old aged patients.

KEY BOX 19.21

RADIOIODINE THERAPY

- Today there is '**no restriction of age and gender**'. (However, this is preferred in children only after completion of growth and in adults only after family is complete)
- Absolute contraindication is pregnancy
- Conception must be avoided for a period of 4 months after radioiodine therapy

KEY BOX 19.22

SECONDARY THYROTOXICOSIS
EFFECTS ON CVS

- Tachycardia
- Wide pulse pressure
- Systolic scratch (Means-Lerman scratch)[1]
- Extrasystoles
- Atrial fibrillation
- Cardiac failure

[1] Means-Lerman scratch is occasionally heard in left 2nd ICS during expiration due to hyperdynamic circulation in thyrotoxicosis (Described by J. Lerman and J.H Means of Massachusetts General Hospital)

Table 19.7 Summary of toxic goitre

Disease	First line	Treatment
1. Primary thyrotoxic cases	Antithyroid drugs	< 45 years—antithyroid drugs > 45 years—radioiodine or surgery
2. Toxic nodular goitre	Antithyroid drugs	Subtotal/total thyroidectomy
3. Solitary toxic nodule	Antithyroid drugs	> 45 years—radioiodine > 45 years—hemithyroidectomy
4. Recurrent thyrotoxicosis after surgery	Antithyroid drugs	Antithyroid drugs/radioiodine therapy
5. Toxic goitre in pregnancy	Propylthiouracil	1st trimester drugs 2nd/3rd—surgery
6. Toxic goitre in children	Antithyroid drugs	Continue drugs/surgery in selected cases

- Propranolol controls the disease very well.
- Radioiodine therapy is the treatment of choice.

3. ***Hyperthyroidism in pregnancy***
   - Invariably it is Graves' disease.
   - In the first trimester, surgery and radioiodine are contraindicated. Carbimazole and propylthiouracil cross the placenta.
   - Surgery, if necessary, can be done in 2nd trimester.
4. ***Apathetic thyrotoxicosis***
   - Thyrotoxicosis in elderly wherein pulse rate is low, they appear to be hypothyroid rather than hyperthyroid. Thyroid gland is rarely palpable.
   - Lethargy and behaviour changes.

Summary of the toxic goitre (Table 19.7)

NEOPLASTIC GOITRE

Adenoma

- The benign tumours of the thyroid gland are not uncommon. They present as a solitary nodule, thus causing a worry to the clinician. Adenomas are of follicular type.
- The diagnosis is established by histological examination.
- Adenomas are treated by hemithyroidectomy/lobectomy.
- However, FNAC cannot distinguish between a follicular adenoma and follicular carcinoma. Hence, a frozen section has to be done.

MALIGNANT TUMOURS[1]

- Thyroid is the only endocrine gland wherein malignant tumours are easily accessible to clinical examination.
- Thyroid is the only endocrine gland wherein malignant tumours occur in children, young age, middle age, old age, and in both sexes.
- Thyroid is the only endocrine gland wherein malignant tumours spread by all possible routes—local, lymphatic and blood spread.
- Thyroid is the only endocrine gland wherein malignant tumours are usually nonfunctional.

Malignant tumours of the thyroid are common. They are interesting tumours, having good prognosis if diagnosed early. Papillary and follicular carcinoma are well differentiated, medullary carcinoma are poorly differentiated.

PAPILLARY CARCINOMA THYROID (PCT)

Aetiology

1. Irradiation to the neck during childhood: In olden days ***radiotherapy was given for benign conditions such as acne in teenagers or enlarged tonsils or thymus gland. Those children had increased risk of papillary carcinoma thyroid. These indications are obsolete now.*** However, accidental radiation to the neck or radiation given to Hodgkin's lymphoma can precipitate the development of papillary carcinoma thyroid.
2. It can be a complication of Hashimoto's thyroiditis.
3. Papillary cancer of thyroid occurs more often in patients with Cowden's syndrome, Gardner's syndrome or Carney's syndrome.
4. Associated mutations: Chromosomal translocation involving RET proto-oncogene (tyrosine kinase) chromosome 10q11.

PEARLS OF WISDOM

Neck radiation increases risk not only for carcinoma thyroid but also for parotid gland tumours and hyperparathyroidism.

Pathology

- It is made up of colloid-filled follicles with papillary projections. In some cases, calcific lesions are found which are called ***psammoma bodies.*** These are diagnostic of papillary carcinoma of thyroid. Characteristic pale, empty, nuclei are present in a few cases which are described as

[1]These were the teachings or sayings of Late Prof Sharath Chandra, FRCS, Professor of Surgery, Madras Medical College, Chennai (Conveyed to me by Late Prof. Subbu, MMC, Chennai, when for the first and last time, I sat with Prof. Subbu as a co-examiner at Govt. Medical College, Coimbatore, 1998).

Orphan Annie eye nuclei. Thyroid gland has very rich intrathyroidal lymphatic plexus. Papillary carcinoma can be unifocal or multifocal (Figs 19.25 and 19.26).

- **Papillary microcarcinoma:** They measure 1 cm or less in diameter. Distant metastasis is extremely rare. **Hence a simple hemithyroidectomy is the treatment of choice.**
- **Follicular variant of papillary cancer:** This is a mixed lesion with a predominance of follicles over papillae. These are treated by total thyroidectomy. It is called **Lindsay tumour.**
- **Tall cell papillary cancer:** This is an aggressive and rapidly growing tumour. It occurs in elderly patients and should be treated by total thyroidectomy.
- Other details are given in Key Box 19.23.

Clinical presentation

- Young females are commonly affected (in the age of 20–40 years).
- It can present as a solitary nodule.
- Very often, the lymph nodes in the lower deep cervical region are involved and thyroid may or may not be palpable.

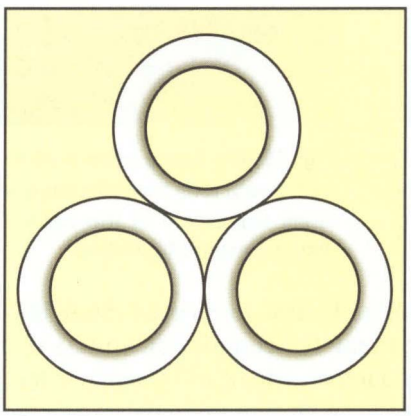


Fig. 19.25: Orphan Annie-eyed nuclei

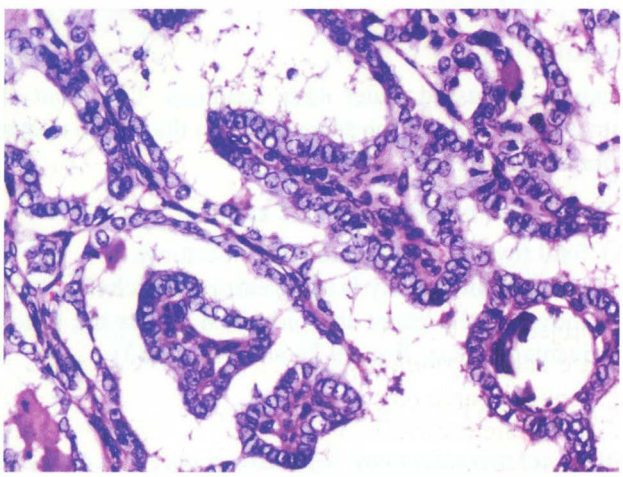


Fig. 19.26: PCT showing papillae with fibrovascular cores lined by follicular cells with Orphan Annie nucleus (*Courtesy:* Dr Laxmi Rao, Head of the Department, Pathology, KMC, Manipal)

KEY BOX 19.23

PATHOLOGY OF PAPILLARY CARCINOMA MICROSCOPY

- **C**alcification
- **C**ystic changes/necrosis
- **C**uboidal pale cell with grooving
- **C**rowded nuclei
- **C**ytoplasmic inclusions—intranuclear
- **C**artoon character—Orphan Annie
- **C**alcified deposits—Psammoma bodies

Observe **7 Cs**

Types—Woolner classification

- Minimal/Occult/Microcarcinoma: Tumour of 1 cm or less in size with no invasion/no lymph node metastasis
- Intrathyroidal: Confined to thyroid gland
- Extrathyroidal: Invasion of adjacent structures.

When thyroid gland is not palpable, it is called occult (hidden). However, papillary carcinoma less than 1.5 cm in diameter is also called 'occult' (Key Box 19.24).

- A few patients present late to the hospital with fixed nodes in the neck, and fixed thyroid to the trachea with or without recurrent laryngeal nerve paralysis (Figs 19.27 to 19.32).

PEARLS OF WISDOM

'It should be noted that papillary carcinoma can be offered as a clinical diagnosis only in the presence of significant lymph nodes in the neck along with a thyroid swelling.'

KEY BOX 19.24

PAPILLARY CARCINOMA—LYMPH NODE METASTASIS—PECULIARITIES

1. They may be palpable even when thyroid gland is not palpable—occult primary
2. Very slow growing
3. Very often, they are intracapsular
4. They need not be hard, are often cystic and firm in consistency
5. At operation, they are bluish in colour because of rupture of the papillae
6. Presence of lymph node metastasis does not affect the prognosis
7. Mostly central neck nodes are cleared. Dissection of posterior triangle and suprahyoid dissection is not necessary.

Prognostic criteria

There are many prognostic criteria that have been used in cases of well-differentiated carcinoma. Various scoring systems are available for well-differentiated carcinomas. Some examples are given in the next page.

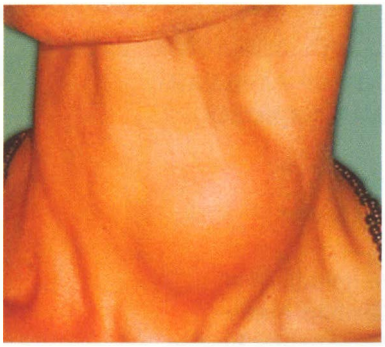


Fig. 19.27: Papillary carcinoma thyroid presenting as solitary nodule. FNAC gave the correct diagnosis. Total thyroidectomy was done in this patient

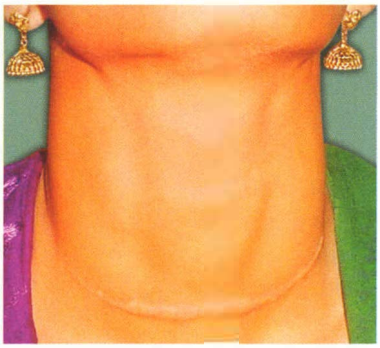


Fig. 19.28: This lady underwent total thyroidectomy for MNG. It was reported as PCT. After one year she presented with a solitary lymph node

Fig. 19.29: This lady underwent hemithyroidectomy two years back and came with recurrence in the opposite lobe. Completion thyroidectomy was done

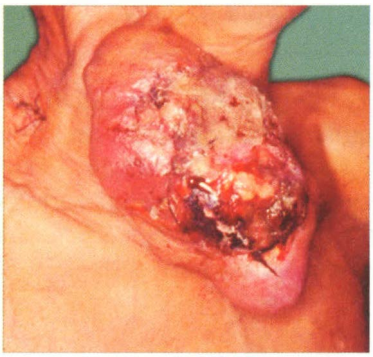


Fig. 19.30: Papillary carcinoma thyroid in a 75-year-old lady—it behaves aggressively in elderly patients. Total thyroidectomy was done (uncommon presentation)

Fig. 19.31: Another case of papillary carcinoma thyroid with huge lymph node secondaries—advanced, neglected case

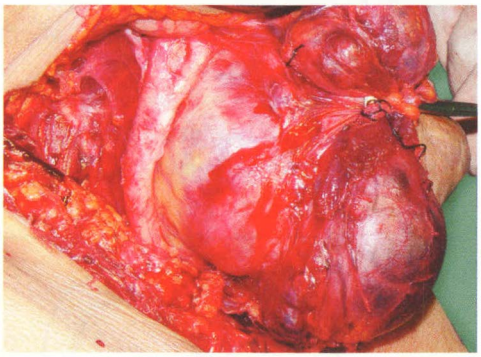


Fig. 19.32: Same patient as in Fig. 19.31—underwent total thyroidectomy with functional block dissection of the neck. *See also* the displacement of carotids

AMES scoring
- **A:** Age less than 40 years—better prognosis
- **M:** Distant metastasis—poor prognosis
- **E:** Extent of tumour extracapsular spread—poor prognosis
- **S:** Size less than 4 cm, good prognosis

AGES scoring
- **A:** Age less than 40 years better prognosis
- **G:** Grade of the tumour—high grade—poor prognosis
- **E:** Extracapsular spread—poor prognosis
- **S:** Size less than 4 cm, good prognosis

MACIS scoring
- **M:** Metastasis
- **A:** Age
- **C:** Completeness of resection
- **I:** Invasion
- **S:** Size

Histological surprise

If a patient undergoes hemithyroidectomy for suspicious adenoma and histopathology reported is papillary carcinoma, dilemma exists as to how to proceed. Straightaway, one can advice the patient for completion (total) thyroidectomy. According to another school of thought, if the patient is in low risk category with intrathyroidal malignancy and other factors favouring good prognosis, then 'wait and watch' policy can be undertaken.

Treatment

It can be discussed under three headings. Treatment of the primary, treatment of the secondaries in the lymph nodes and suppression of TSH.

I. Treatment of the primary/2 choices

(A) Total thyroidectomy and (B) Lobectomy.

A. Total thyroidectomy is the treatment of choice. It means removal of the entire thyroid gland. It has the following advantages (Key Box 19.25 and Fig. 19.33).

PEARLS OF WISDOM

After total thyroidectomy, thyroxine is not given for a period of 4 weeks so that thyroid remnants can be ablated with radioiodine—till TSH are levels are above 30 MIU/L

KEY BOX 19.25

PAPILLARY CARCINOMA
TOTAL THYROIDECTOMY ADVANTAGES

1. Easy to detect and treat residual or metastatic disease.
2. Easy to assess recurrence by thyroglobulin level estimation.
3. Eliminates contralateral occult cancer (multifocal in 80% patients)
4. Eliminates resurgery.
5. Eliminates risk of recurrence, thus improving survival.
6. TSH suppression can be done.

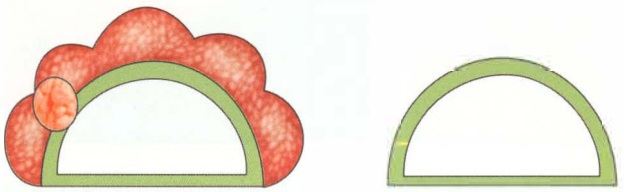


Fig. 19.33: Total thyroidectomy—in majority of cases

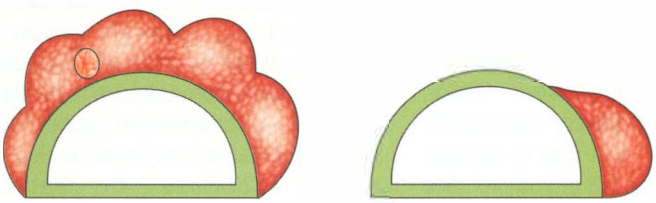


Fig. 19.34: Lobectomy—low risk groups (hemithyroidectomy)

Dose of radioiodine 30–100 mCi.

B. Lobectomy (hemithyroidectomy) means removal of one lobe and entire isthmus. Those who favour lobectomy have following arguments (Fig. 19.34):

1. Total thyroidectomy has high complication rates such as RLN paralysis, permanent hypothyroidism, permanent hypoparathyroidism, etc.
2. Recurrence in opposite lobe is only 5% which means in 95% of the cases, removal of the opposite lobe is unnecessary.
3. Even if it occurs later, since it is not dissected at the time of initial surgery, the lobe can be removed easily.
4. **Tumour multicentricity has little prognostic significance.** Thus in a few selected cases, lobectomy can be done.

Advantages of lobectomy

1. No hormone replacement
2. No hypoparathyroidism
3. Need not test thyroid function regularly.

II. Treatment of secondaries in the lymph nodes

- Mostly central neck nodes are cleared. If nodes are enlarged in the anterior triangle, they are dissected and removed *en bloc* along with fat and fascia. This is called **functional block dissection** (**Berry picking** means removal of enlarged lymph nodes only. It is no longer followed).
- Structures such as **internal jugular vein, sternomastoid muscle, accessory nerve are not removed** because lymph nodes are slow growing and they rarely spread/ outside the capsule of the lymph node. However, in exceptional cases of papillary carcinoma with infiltration to these structures, there should not be hesitation to remove these structures.
- **Lateral aberrant thyroid:** Initially believed to be thyroid tissue. It is actually metastasis into levels 3 and 4 lymph nodes from papillary carcinoma thyroid.
- **No role for prophylactic neck dissection except in children**
- *See* Figs 19.35 to 19.38 for various pictures of papillary carcinoma thyroid.

III. Suppression of the TSH

This is an important aspect in the postoperative period because papillary carcinoma is a TSH dependent tumour. To prevent the patient developing hypothyroidism in the postoperative period and to suppress TSH, thyroxine 0.3 mg/ day is given.

- Failure of suppression of TSH to a level < 0.1 mU/litre suggest inadequate dose of thyroxine or noncompliance.

When to give tri-iodothyronine (T3) in the follow-up period of well-differentiated thyroid cancers?

- Patients who require regular radio-iodine for scanning and ablation should be given T3 because it acts quickly and it can be stopped and restarted quickly. On the other hand, T4 has to be stopped for almost 30 days prior to scanning and ablation rendering patients severely hypothyroid for 4 weeks. (Dose of T3 is 40–60 μg/day. It is very costly; not freely available).

Management protocol of papillary carcinoma of the thyroid (Fig. 19.39)

Summary of papillary carcinoma thyroid as shown in Key Box 19.26.

KEY BOX 19.26

SUMMARY OF PAPILLARY CARCINOMA THYROID

- Most common histological type of thyroid cancer (60 to 65%)
- Most often it is multifocal (80%)
- Commonly it spreads by lymphatic spread.
- Psammoma bodies are diagnostic of papillary carcinoma of thyroid.
- Most often it presents as solitary nodule and with or without lymph node metastasis
- Most commonly done procedure for papillary carcinoma thyroid is total thyroidectomy with or without functional neck dissection
- Most of the patients (>95%) have 10-year survival rate.

Fig. 19.35: This patient had undergone hemithyroidectomy in a peripheral hospital 15 years back. He did not receive any medication in these years. He presented with ulcerated nodules on the right side of the neck (lymph nodal mass). He successfully underwent completion thyroidectomy with functional neck dissection on the right side. Also observe sebaceous cyst on the right side of the chest wall (*Courtesy:* Dr Annappa Kudva, Prof of Surgery, KMC, Manipal)

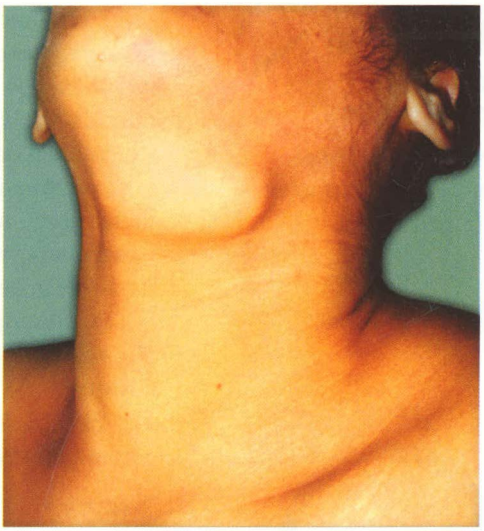


Fig. 19. 36: This 24-year-old lady presented with swelling in the submandibular region. FNAC revealed salivary tissue. Hence, submandibular sialoadenitis was diagnosed. At surgery, the salivary gland was separate from this mass which was excised. Histopathology reported as papillary carcinoma arising from thyroglossal duct cyst in ectopic area. She is on L-thyroxine 0.3 mg/day for last 18 months. As of now she has no complications. (*Courtesy:* Dr Padmanabha Bhat, Prof of Surgery, KMC, Manipal)

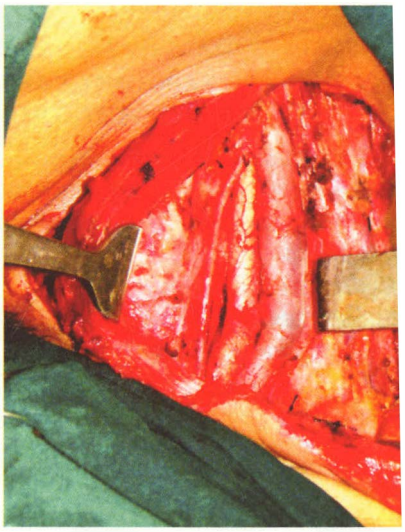


Fig. 19.37: Functional neck dissection is done for papillary carcinoma thyroid. Routine central compartment dissection is not required. Figure shows vagus nerve, carotid artery and internal jugular vein. Sternocleidomastoid is retracted. All these structures are preserved. Hence, the name functional neck dissection (*Courtesy:* Dr Gabriel Rodrigues, Prof of Surgery, Dr Suresh BP, Asst Professor, Dept. of Surgery, KMC, Manipal)

TNM STAGING IS GIVEN IN NEXT PAGE

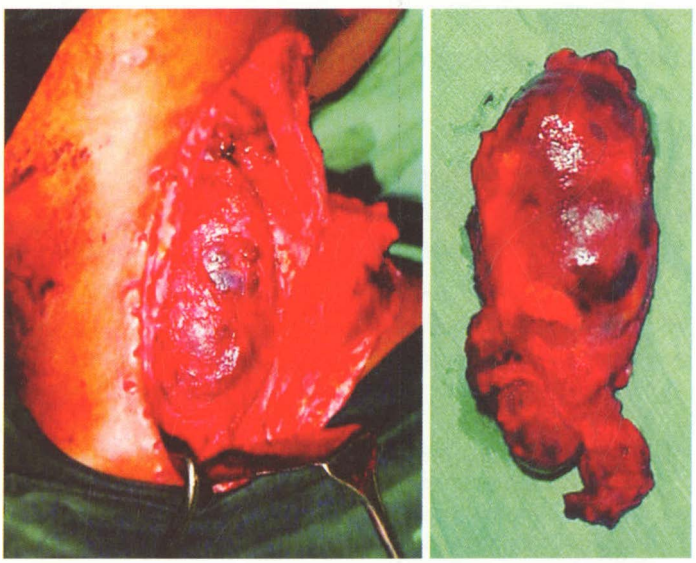


Fig. 19.38: Observe bluish colour lymph nodes at surgery and specimen of lymph nodes

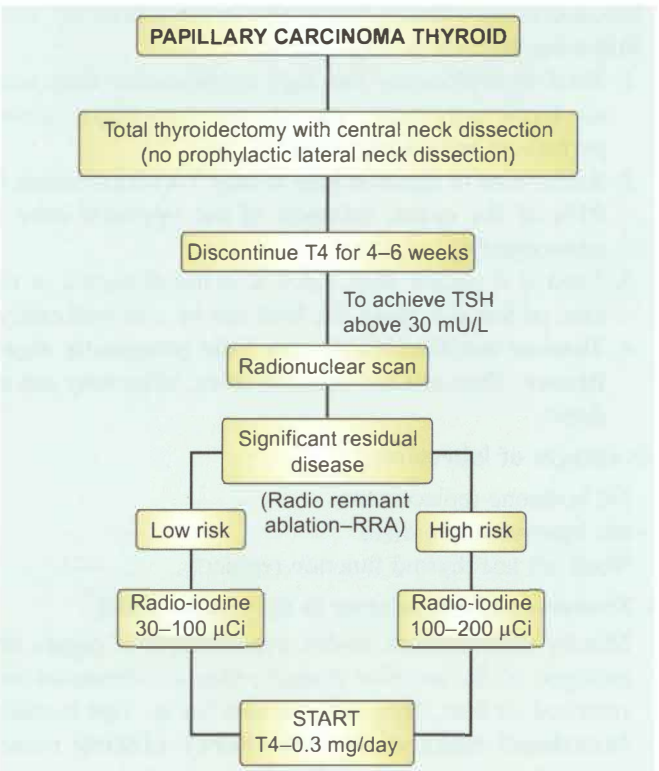


Fig. 19.39: Management protocol of papillary carcinoma

TNM STAGING Thyroid cancer

Tumour
Tx : Primary cannot be assessed
T0 : No evidence of primary
T1 : Limited to thyroid, 2 cm or less
T2 : Limited to thyroid > 2 cm but < 4 cm
T3 : Limited to thyroid > 4 cm
T4 : Extending beyond capsule any size

Nodes
Nx : Cannot be assessed
N0 : No regional node metastasis
N1 : Regional node metastasis

Anaplastic carcinoma is always stage IV

Metastases
Mx : Cannot be assessed
M0 : No metastasis
M1 : Metastasis is present

Separate stage groupings are recommended for papillary or follicular (differentiated), medullary, and anaplastic (undifferentiated) carcinoma

Papillary and follicular thyroid cancer (age < 45 years):

Stage	T	N	M
I	Any T	Any N	M0
II	Any T	Any N	M1

Papillary and follicular; differentiated (age ≥ 45 years):

Stage	T	N	M
I	T1	N0	M0
II	T2	N0	M0
III	T3	N0	M0
IVA	T1–T3	N1a	M0
	T4a	N1b	M0
IVB	T4b	Any N	M0
IVC	Any T	Any N	M1

Anaplastic carcinoma (all anaplastic carcinomas are considered stage IV):

Stage	T	N	M
IVA	T4a	Any N	M0
IVB	T4b	Any N	M0
IVC	Any T	Any N	M1

Medullary carcinoma (all age groups):

Stage	T	N	M
I	T1	N0	M0
II	T2, T3	N0	M0
III	T1–T3	N1a	M0
IVA	T4a	N0	M0
	T4a	N1a	M0
	T1	N1b	M0
	T2	N1b	M0
	T3	N1b	M0
	T4a	N1b	M0
	T4a	N0, N1b	M0
	T1–T4a	N1b	M0
IVB	T4b	Any N	M0
IVC	Any T	Any N	M1

FOLLICULAR CARCINOMA

- ***Incidence:*** Constitutes 17% of cases (Key Box 19.27).
- Follicular adenoma 20% are malignant and 80% are benign

Aetiology

- Follicular carcinoma usually arises in a multinodular goitre, especially in cases of endemic goitre. It should be suspected when MNG starts growing rapidly.
- It can present as solitary nodule also (Figs 19.40 and 19.41).

KEY BOX 19.27

INCIDENCE OF THYROID MALIGNANCY

• Papillary carcinoma	60–65%
• Follicular carcinoma	15–20%
• Anaplastic carcinoma	10–12%
• Medullary carcinoma	5–10%
• Others	10%

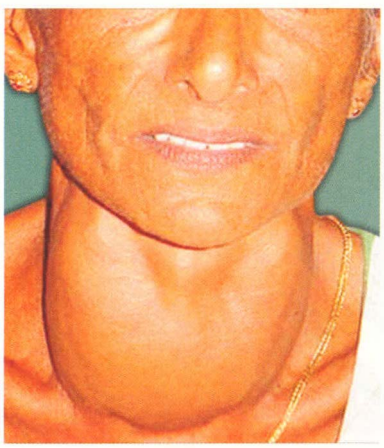


Fig. 19.40: Follicular carcinoma thyroid—clinically suspected—ultrasound showed microcalcification—FNAC revealed follicular cells. She underwent total thyroidectomy. Final report was follicular carcinoma thyroid

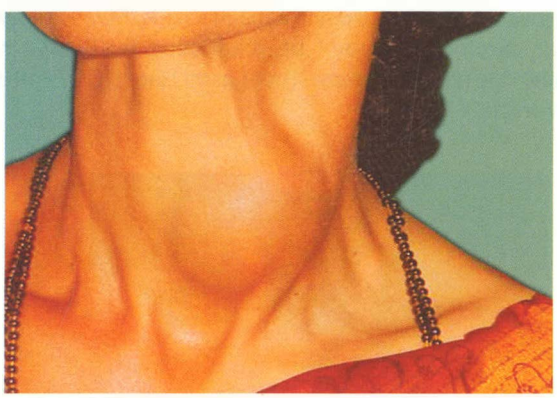


Fig. 19.41: A 35-year-old female with solitary nodule. FNAC revealed papillary carcinoma

Pathology

Depending upon the property of invasion it is classified into:
- **Noninvasive** which means minimal invasion.
- **Invasive** refers to angio-invasion and capsular invasion, necessary for the diagnosis of follicular carcinoma of

thyroid. The tumour cells line the blood vessels and get dislodged into the systemic circulation producing secondaries in the bones. Microscopically, most of the tumours are well-encapsulated (Fig. 19.42).

Clinical presentation (Figs 19.49 to 52)

- It can present as a solitary nodule. The diagnosis is considered only after an ultrasound scan reveals some features of malignancy such as **microcalcification**. Peak age group is around 40 years (Fig. 19.43).
- In case of long-standing multinodular goitres, if the goitre is rapidly growing, hard or has restricted mobility, follicular carcinoma can be considered.
- **Metastasis in the flat bones:** The only clinical situation wherein a follicular carcinoma can be considered as the diagnosis is when a patient with a thyroid swelling presents with metastasis in the bone in the form of bony swelling or pathological fractures. Commonly, secondaries develop in the flat bones such as skull, ribs, sternum, vertebral column because the flat bones retain red marrow for a longer time. **When bony swelling is obvious and thyroid is not palpable clinically, it is called occult primary site.**
- **The clinical features of secondary in the skull are:**
  - They are rapidly growing
  - They are warm
  - Vascular and pulsatile
  - Underlying bony erosion may be present (Figs 19.44 to 19.52 and clinical notes).

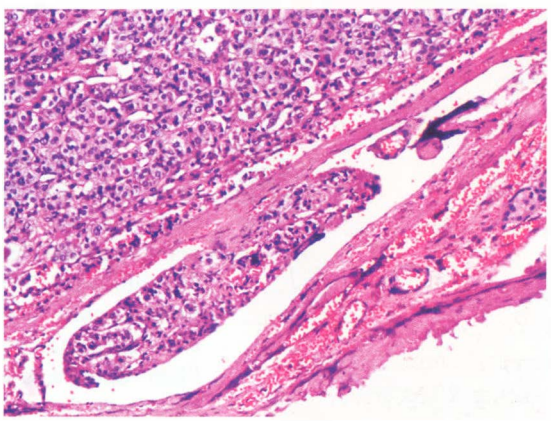


Fig. 19.42: Follicular carcinoma thyroid showing well differentiated follicles invading the capsule and capsular veins. Please remember follicular carcinoma cannot be diagnosed by FNAC but by histopathology only (*Courtesy:* Prof Laxmi Rao, HOD, Pathology, KMC, Manipal)

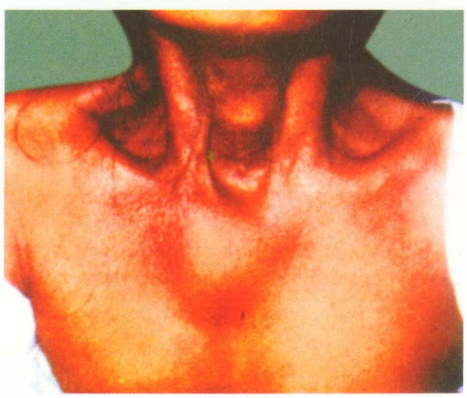


Fig. 19.43: Secondary deposit in the sternum in a patient who underwent near-total thyroidectomy for follicular carcinoma 5 years back

Fig. 19.44: Chest X-ray showing osteolytic lesion in the left clavicle

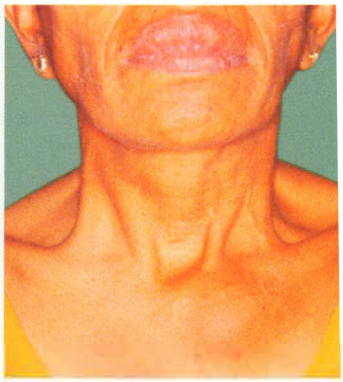


Fig. 19.45: This lady presented with secondary in the left second rib. She did not have thyroid swelling. A case of occult follicular carcinoma

Fig. 19.46: Carefully watch right sternoclavicular joint—she had a tender bony swelling

CLINICAL NOTES

A patient with a diagnosis of "lipoma of the scalp" was posted for excision in the prone position. As the chief surgeon scrubbed and was about to paint the part, he could see the pulsatile nature of the swelling, which he had missed in the outpatient department. Fortunately, it was not excised. A needle was introduced and frank blood was aspirated. Surgery was cancelled. X-ray skull showed osteolytic lesion. She had a small thyroid nodule.

Investigations

- Routine investigations
- **Ultrasound scan** is done (Figs 19.53 and 19.54) to demonstrate nature of the nodule, whether solid or cystic and to guide FNAC.
- **FNAC of the nodule.** It should be remembered that FNAC cannot differentiate a follicular adenoma from follicular carcinoma. Hence, if FNAC reports as follicular cells, **overtreat the patient** by total thyroidectomy. Some follow with frozen section and proceed. This is not favoured by many. If it is reported as follicular carcinoma, no further surgery is required. If it is reported as benign, patient requires to be treated for hypothyroidism. **Open biopsy is not taken in operable cases** (Key Box 19.28).

KEY BOX 19.28

MALIGNANT TUMOURS WHEREIN OPEN BIOPSY IS NOT TAKEN

- Thyroid
- Parotid
- Testis

CLINICAL NOTES

A 38-year-old lady underwent total thyroidectomy for a solitary nodule, with FNAC revealing follicular carcinoma. After3 years, she came with pain on the right side of sternum. Careful examination revealed a tender, warm, bony swelling arising from sternoclavicular joint. The diagnosis was follicular carcinoma of the thyroid. No wonder it is said that a surgeon should have eagle's eyes! (Fig.19.48)

MS exam case 2008, JNMC—Belgaum, Contributed by Prof. Ashok Godhi, HOD of Surgery

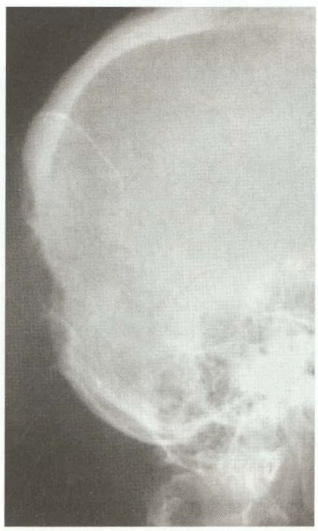


Fig.19.47: Erosion of skull bone

KEY BOX 19.29

CAUSES OF SECONDARY IN THE SKULL

1. Follicular carcinoma of thyroid
2. Renal cell carcinoma
3. Hepatocellular carcinoma
4. Prostatic carcinoma
5. Bronchogenic carcinoma

- **CT scan** in appropriate cases (Fig.19.55).
- **Alkaline phosphatase**—if increased, bone scan should be done.
- **Plain X-ray** of the involved bone can reveal **osteolytic lesions** (Fig. 19.48 and Key Box 19.29).
- When primary is not found, **bone biopsy** is required to find out the site of the primary.

Treatment of follicular carcinoma of thyroid

I. Treatment of the primary

- ***Situation I:*** When a patient has enlarged thyroid gland and scalp swelling, total thyroidectomy is the treatment of choice. Secondaries do not take up the radioisotope (^{131}I) in the presence of primary tumour. Hence, lobectomy or hemithyroidectomy should not be done.
- ***Situation II:*** A patient undergoes subtotal thyroidectomy for MNG and final report is follicular carcinoma thyroid. In such cases, it is better to do **completion thyroidectomy within 7 days** or **after 4 weeks.**
- If done, within 7 days, or after 4 weeks it is relatively easy to do. Between 7 days and 4 weeks, the inflammatory process would have produced dense fibrosis, chances of injuring RLN, parathyroids are high.
- ***Situation III:*** A patient with solitary nodule—high suspicion of malignancy after sonography and FNAC reported as follicular cells, it is better to go ahead with total thyroidectomy.

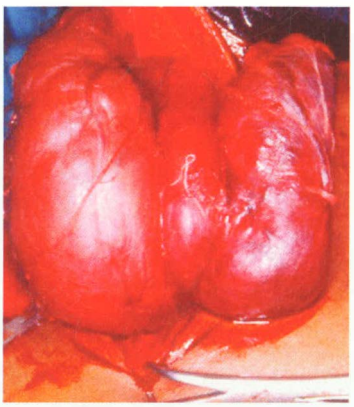


Fig. 19.48: Total thyroidectomy in progress. This is the best option for most of Indian patients who present with fairly big lesions. The need to take a small dose of thyroid hormone lifelong is a disadvantage but if the report is 'malignancy', it is the treatment

FOLLICULAR CARCINOMA THYROID—INTERESTING PICTURES

Figs 19.49 and 19.50: This lady presented with thyroid swelling of 5 years' duration with swelling in the scalp and sternum of 2 months duration. Bony swellings were painful—it was a case of follicular carcinoma of thyroid with multiple bony metastasis: **Lady with three swellings**

Fig. 19.51: Ulcerated secondary in the scalp bone from follicular carcinoma thyroid. See dilated veins in the neck. No other differential diagnosis in such patients

Fig. 19.52: This lady, who had undergone hemithyroidectomy for follicular carcinoma thyroid 3 years back, presented with recurrence. Total thyroidectomy could be done

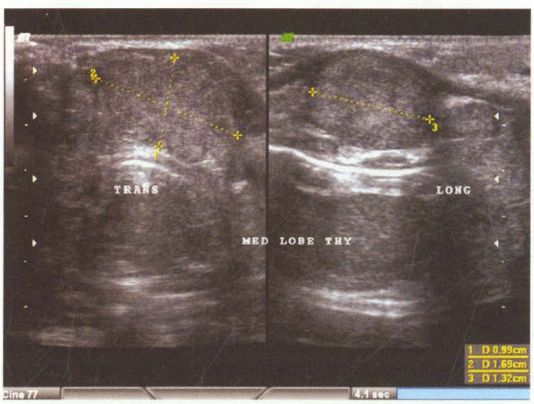


Fig. 19.53: Ultrasound revealing recurrence (nodules) in both lobes of the thyroid gland (*Courtesy:* Dr Chandrakanth Shetty, Prof of Radiodiagnosis, KMC, Manipal)

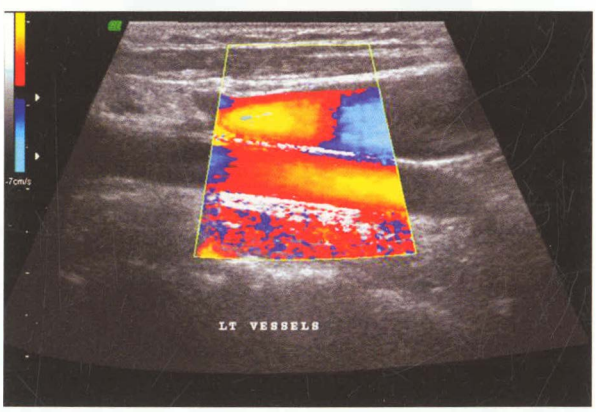


Fig. 19.54: Colour Doppler showing the relationship of the gland to major vessels of the neck (*Courtesy:* Dr Chandrakanth Shetty, Professor of Radiodiagnosis, KMC, Manipal)

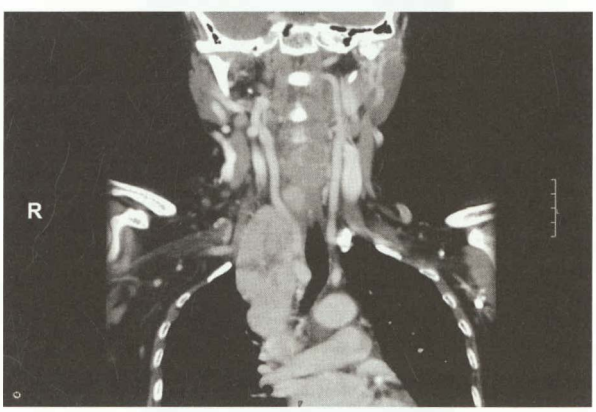


Fig. 19.55: CT neck and chest showing the large mass displacing the carotid artery, internal jugular vein thrombosis and mediastinal lymph nodes. Lymph nodes were cleared after sternotomy

II. Treatment of the metastasis

After total thyroidectomy, a whole body bone scan is done to look for metastasis in the bone. A single secondary can be treated by oral radioiodine therapy, followed if necessary by external radiotherapy depending upon the response of the tumour. Multiple secondaries, if present, are treated by oral radioiodine therapy.

III. Postoperative thyroxine

- In the postoperative period, patients should receive thyroxine 0.3 mg/day to suppress TSH and to supplement thyroxine.
  Prognosis: 15% mortality in 10 years.

HÜRTHLE CELL CARCINOMA

- Hürthle cell carcinoma is a **variant of follicular carcinoma** (Key Box 19.30). It is more aggressive than follicular carcinoma.
- These tumours are defined by the presence of more than **75% of follicular cells** having oncocytic features.
- Tumour contains sheets of eosinophilic cells packed with mitochondria.
- **They secrete thyroglobulin.**
- Even if Hürthle cell adenoma is well encapsulated, it is **potentially malignant**.
- It does not take up ^{131}I. Hence, it is less likely to respond to ^{131}I ablation.
- Higher mortality (20% at 10 years).
- 99mTc—sestamibi scan can detect Hürthle cell carcinoma.

Criteria to diagnose Hürthle cell carcinoma

- Capsular/vascular invasion, distant metastasis.
- Higher chance of spread to lymph nodes compared to follicular thyroid carcinoma.
- Higher chances of spread to distant sites also.

Treatment

- Total thyroidectomy is the treatment of choice. In many cases of Hürthle cell carcinoma, lymph nodes are enlarged. Hence, modified radical neck dissection is done (MRND).
- TSH suppression and follow-up are regularly required.

PEARLS OF WISDOM

All Hürthle cell neoplasms are almost malignant and all adenomas are almost follicular

In all suspicious solitary nodules of follicular cell origin, it is better to overtreat them by total thyroidectomy than relying upon the frozen section, to avoid second surgery.

Follow-up of patients with papillary and follicular carcinoma thyroid—differentiated thyroid cancer

- **Serum thyroglobulin (Tg):** Thyroid is the only organ which produces thyroglobulin. Levels greater than 1 to 2 mg/ml in patients receiving replacement thyroxine therapy indicates presence of metastasis. Hence, assess the serum Tg response to injected recombinant human TSH, every year. In thyrotoxicosis and thyroiditis, thyroglobulin levels may be increased (Key Box 19.31).

KEY BOX 19.30

HÜRTHLE CELL CARCINOMA

- More aggressive follicular carcinoma
- More than 75% malignant cells
- More chances of lymphatic spread
- More chances of distant spread
- More chances of mortality

KEY BOX 19.31

SERUM THYROGLOBULIN

- Normal value: < 1–35 µgm/litre
- Above 50 µgm/litre suggest malignancy
- Above 100 µgm/litre suggest pulmonary or skeletal metastasis
- It is produced by follicular cells of the thyroid. Hence elevated in well-differentiated carcinoma. **Not in medullary carcinoma thyroid**.
- Hence it is a tumour marker of well-differentiated carcinoma
- Anti-thyroglobulin antibodies (ATA) TgAb are found in Hashimoto's thyroiditis and in Graves' disease

- **Ultrasonography or MRI** scans of the neck for localisation of residual or recurrent tumour.

ANAPLASTIC CARCINOMA

Incidence 10–12% of cases

Clinical features (Key Box 19.32)

- Common in elderly women around 60–70 years of age.
- Majority of the patients present with rapidly-growing thyroid swelling of short duration. The surface is **irregular** and consistency is **hard.**
- **Early infiltration of the trachea** results in stridor (scabbard trachea).

KEY BOX 19.32

ANAPLASTIC CARCINOMA THYROID

- The most rapidly growing thyroid malignancy
- Advanced age group at presentation
- Advanced nature of presentation
- Gross local infiltration—Berry's sign positive
- No form of treatment is successful
- Intrinsic carcinoma of larynx spreading outside and infiltrating the skin should be considered as a differential diagnosis

- Infiltration of carotid sheath: In such cases, common carotid artery pulsation will not be palpable. This is described as **'Berry sign positive'.**
- **Early fixity** is characteristic. Thus, the resectability rate is almost nil (Figs 19.56 and 19.57).

Diagnosis

It is established by FNAC, other investigation being CT scan (Fig. 19.58).

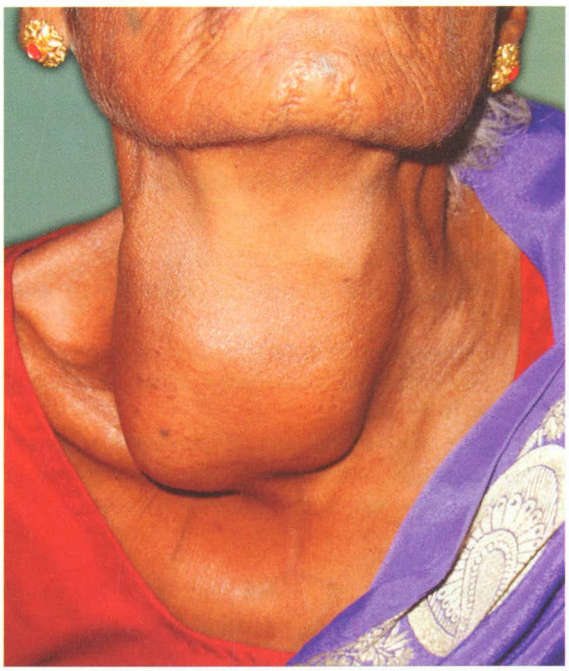


Fig. 19.56: Hard nodular fixed thyroid of three months duration. FNAC—anaplastic carcinoma

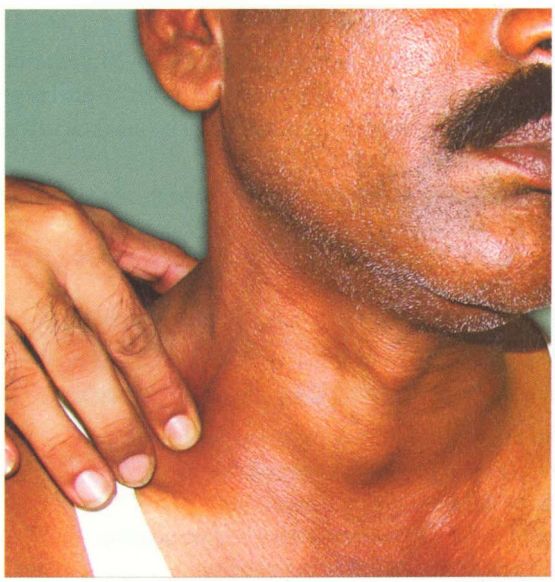


Fig. 19.57: A 40-year-old male with 3 cm hard, fixed nodule with recurrent laryngeal nerve paralysis. FNAC—anaplastic carcinoma

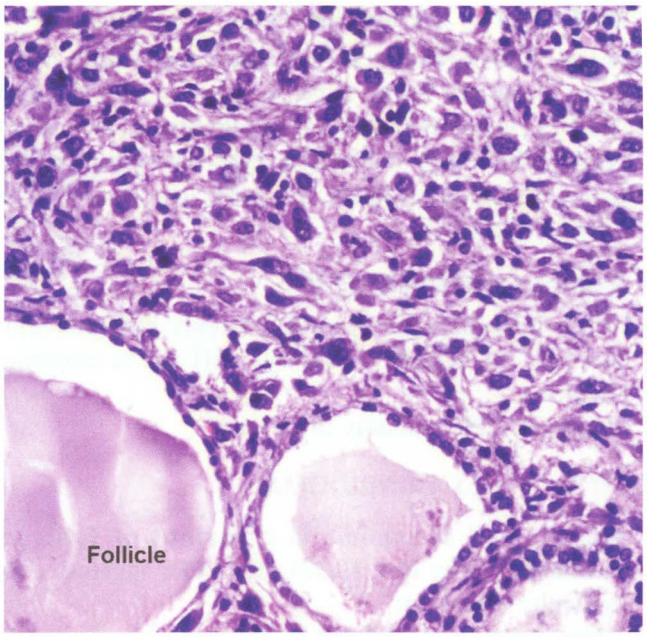


Fig. 19.58: Highly pleomorphic malignant cells. (*Courtesy:* Pr Laxmi Rao, HOD, Department of Pathology, KMC, Manipal)

Treatment

- Due to the gross local infiltration into the vital structure in the neck such as common carotid artery and trachea the resectability rate is low.
- However, very rarely a surgeon will get an opportunity t excise isthmus so as to relieve compression of the trachea
- Postoperative radiotherapy is given as a palliativ treatment.
- In many cases, death occurs within 6 to 8 months.

MEDULLARY CARCINOMA OF THE THYROID (MCT)

- These tumours arise from parafollicular 'C' cells which are derived from ultimobranchial bodies and not from thyroic follicle.
- These tumours present in two different ways.
  1. Sporadic is common, seen in about 80–90% of cases.
  2. Familial variety present as a part of multiple endocrine neoplasia (MEN).

MEN Type I

- Pituitary adenoma
- Parathyroid adenoma
- Pancreatic adenoma

MEN Type IIa

- Parathyroid adenoma
- Phaeochromocytoma
- Medullary carcinoma of thyroid

- When it is associated with mucocutaneous neuromas involving lips, tongue, eyelids, it is called ***Sipple syndrome,*** with an occasional marfanoid habitus (MEN type IIb) (Fig. 19.59).
- It has got a characteristic ***amyloid stroma*** (Fig. 19.60).
- These tumours are not TSH-dependent and do not take up radioactive iodine (Key Box 19.33).

Hormones produced by MCT

- Calcitonin (Key Box 19.34)
- Prostaglandins
- Serotonin (5-HT), ACTH

Spread

- Both by lymphatics and blood, thus, worsening the prognosis.

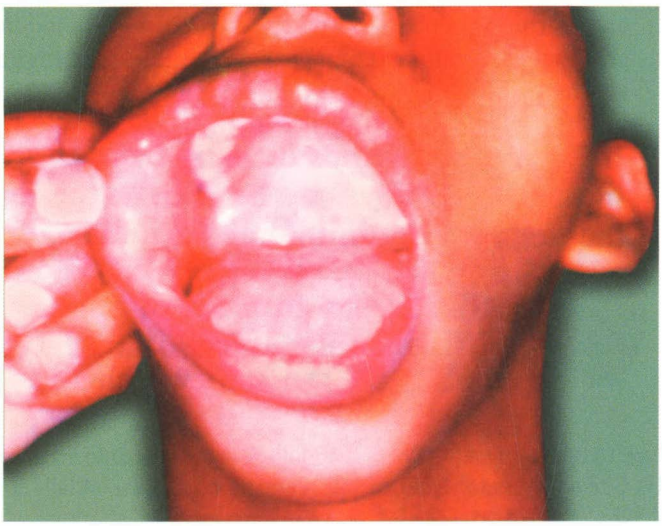


Fig. 19.59: FNAC proved medullary carcinoma with mucocutaneous neuroma—MEN type IIb

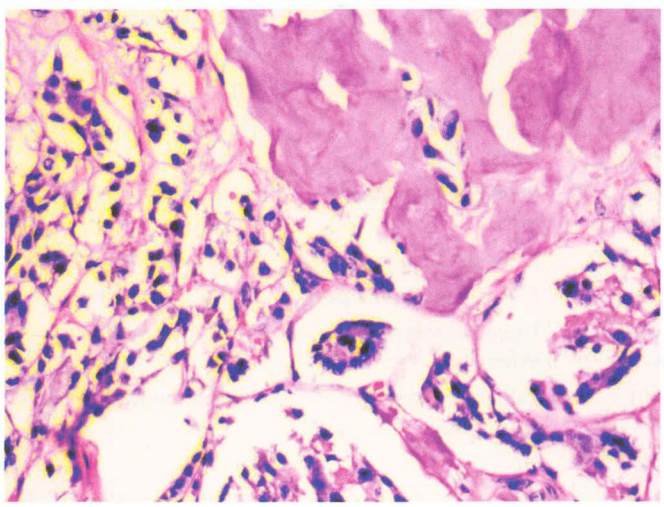


Fig. 19.60: Medullary carcinoma thyroid—characteristic amyloid stroma

KEY BOX 19.33

HOW MCT DIFFERS FROM OTHER DIFFERENTIATED THYROID CANCERS

- It arises from parafollicular cells.
- It can be associated with familial variety.
- Spreads both by lymphatics and blood.
- It produces hormones such as calcitonin and carcino-embryonic antigen.
- Tumour is not TSH-dependent and hence does not take up radioactive iodine.
- Radical neck dissection for lymph node metastasis should be done.
- Prophylactic total thyroidectomy for infants with genetic trait (RET gene mutations).
- Cannot predict prognosis (variable).

KEY BOX 19.34

CALCITONIN AND MEDULLARY CARCINOMA THYROID

- Not measurable in normal persons
- MCT produces very high levels
- It is the tumour marker of MCT
- Level decreases after thyroidectomy
- Level increases in case of recurrence
- Prophylactic thyroidectomy in relatives if calcitonin levels are high.

Treatment

1. Total thyroidectomy with radical neck dissection
   - Before proceeding with surgery, **look for an associated phaeochromocytoma.**
2. The lymph nodes are treated by radical block dissection because they are fast-growing, when compared to papillary carcinoma.

PEARLS OF WISDOM

If there are multiple secondaries in the bone, oral ^{131}I has no role because this tumour does not arise from thyroid cells. Only palliative radiotherapy can be given.

Clinical criteria for the diagnosis of carcinoma of thyroid

1. A thyroid swelling which is rapidly growing.
2. Thyroid swelling with lower deep cervical lymph nodes and lymph nodes in the posterior triangle (papillary carcinoma thyroid) involved.
3. Hard gland, fixed to the trachea—anaplastic carcinoma of the thyroid.
4. Thyroid swelling with a rapidly growing, vascular, pulsatile swelling, commonly in the skull (follicular carcinoma).

5. Thyroid swelling with hoarseness of the voice indicating infiltration of recurrent laryngeal nerve is a feature of malignancy.
6. Thyroid swelling with Berry sign positive (anaplastic carcinoma of the thyroid)
7. Kocher's test positive may be an indication of infiltration into trachea.

Summary of the malignant tumours of the thyroid gland is shown in Table 19.8.

LYMPHOMA

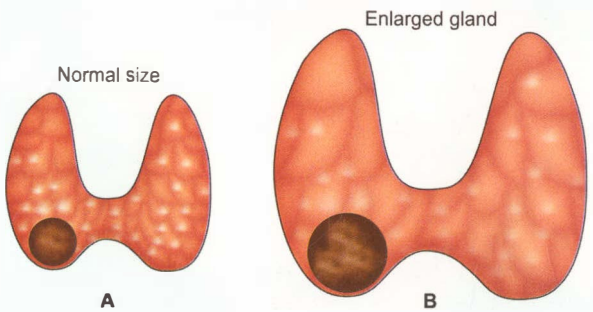
- It is rare. Hashimoto's thyroiditis can predispose to lymphoma.
- Older patients are commonly affected.
- The tumour can present as rapidly-growing, large thyroid swelling (primary lymphoma).
- Sometimes, it can appear as a part of generalised lymphoma (non-Hodgkin's variety).
- FNAC may give the diagnosis—**Tru cut biopsy is ideal.**
- It is interesting to note that lymphomas of the thyroid respond very well to chemotherapy and radiotherapy (Figs 19.61A and B).

Figs 19.61A and B: Lymphoma of the thyroid gland with mediastinal lymph nodes and superior vena caval obstruction. Watch dilated veins and also three cutaneous nodules

SOLITARY NODULE OF THE THYROID GLAND

Definition: Solitary nodule (SN) is a clinical term denoting presence of a single palpable nodule in the thyroid gland **(No other part of thyroid gland is palpable)** (Fig. 19.62A).

- Almost all the thyroid swellings initially can present as a solitary nodule. However, puberty goitres, colloid goitres, diffuse toxic goitres produce uniform enlargement of the thyroid gland. Multinodular goitre (MNG) presents as multiple nodules. However, very often a solitary nodule on clinical examination, may turn out to be a multinodular goitre at exploration. **The solitary nodule has a higher incidence of malignancy when compared to MNG.**



Figs 19.62A and B: (A) Solitary nodule. Rest of the gland is normal (B) Solitary nodule. Rest of the gland is enlarged—mostly it is MNG

Table 19.8 Summary of the malignant tumours of thyroid gland

	Papillary	Follicular	Anaplastic	Medullary
1. Aetiology	Irradiation	Endemic goitre	Unknown	Sporadic or familial
2. Incidence	60%	17%	13%	6%
3. Age (years)	20–40	30–50	50 and above	Middle age
4. Diagnosis	Thyroid swelling with lymph node metastasis	Thyroid swelling with bony metastasis	Thyroid swelling, local fixity, stridor	Difficult to diagnose clinically
5. Microscopy	Orphan Annie eye nuclei, psammoma bodies	Angioinvasion, capsular invasion	Poorly differentiated cells	Amyloid stroma-like carcinoid
6. Spread	Lymphatic	Blood	Local infiltration	Lymphatic, blood
7. Investigation	FNAC	Frozen section	FNAC, incision biopsy	FNAC, calcitonin
8. Treatment of the primary	Near-total/Total thyroidectomy	Near-total/total thyroidectomy	Isthmusectomy, external RT	Total thyroidectomy
9. Treatment of metastasis	Functional neck dissection	Radioiodine ^{131}I or external RT	Palliative external radiotherapy	Radical block dissection
10. TSH dependence	Yes	Yes	No	No
11. Hormone production	Very rare	Very rare	No	Calcitonin, 5-HT, ACTH
12. Prognosis	Excellent	Good	Worst	Bad

- It is a **common surgical problem** encountered by general surgeons. By definition, one can call it a solitary nodule **when only the nodule is clinically palpable** but rest of the gland is not palpable.
- However, when a nodule is palpable and the opposite lobe or any other part of the thyroid is palpable, it is a **'dominant' nodule.** It may probably be a case of multinodular goitre (Fig. 19.62B).

Introduction

- It is the **most common surgical disease** of the thyroid gland (Figs 19.63 to 19.65).
- Incidence—4%
- Sex ratio—4 times more common in women.
- 15–30% patients of SN have nodule on the other side.
- **Solitary solid swelling in a male**—risk of malignancy is 48% but it is ¼ of 48%—(12%) in females.
- **Solitary cystic**—risk of malignancy in males is 24% but in females it is ¼ of this, i.e. 6%.
- In general, incidence of malignancy is 8–10%.
- Ultrasonography followed by FNAC is the first line of investigations.
- Aim is to identify a solitary nodule which has **high risk**, e.g. **male, solid, solitary nodule** (Key Box 19.35).

KEY BOX 19.35

HIGH RISK SOLITARY NODULE

- **Male** patients with **'solid'** nodule
- Rapid growing nodule of short duration
- Nodule which has **restricted** mobility
- Nodule with jugular **lymphadenopathy** on ultrasound
- Nodule which is **hard in consistency**
- Nodule with **hoarseness of voice**

Causes of solitary nodule of the thyroid gland

1. In 50% of the cases, a clinically palpable solitary nodule is a part of multinodular goitre
2. Toxic autonomous nodule
3. Adenoma
4. Carcinoma
5. Cysts.

PEARLS OF WISDOM

About 2 to 3% of solitary nodules are associated with hyperthyroidism. Chances of malignancy in a hyperthyroid nodule is very low.

Indications for surgery

- Malignant cytology
- Suspicious cytology
- Large size
- Pressure effect, substernal extension
- Clinical suspicion of malignancy
- Cosmesis
- Male sex
- Cyst-recurs after aspiration

Investigations

1. Blood investigations

- T3, T4 and TSH: Free thyroxine—Better indicator of thyroid status than total T4. T3 is useful when T4 level is border line (T3 toxicosis). TSH is needed when there is no clinical manifestation of hyper (or) hypothyroidism.
- Thyroid auto-antibodies—when Hashimoto's thyroiditis is suspected.
- Serum thyroglobulin (STG)—tumour marker
  a. Normal value is < 1–35 micrograms per litre

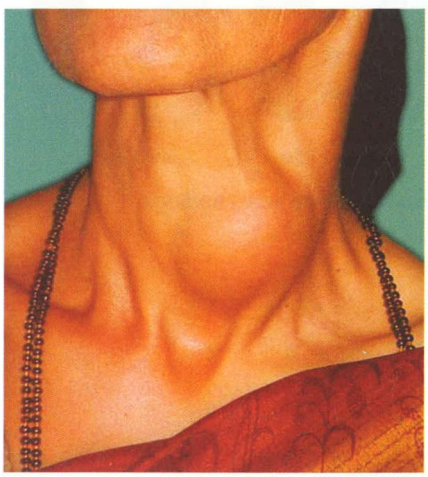


Fig. 19.63: A 35-year-old female with solitary nodule. FNAC revealed papillary carcinoma

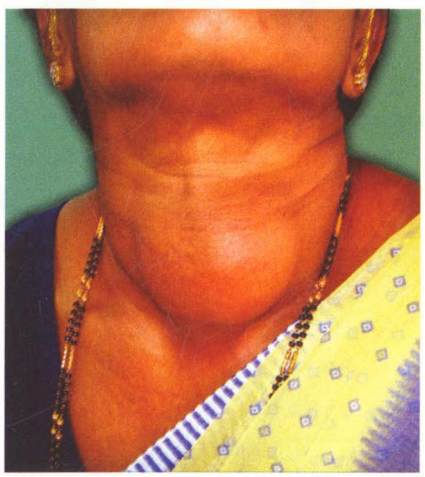


Fig. 19.64: A 45-year-old female with solitary nodule. FNAC revealed thyroid cyst

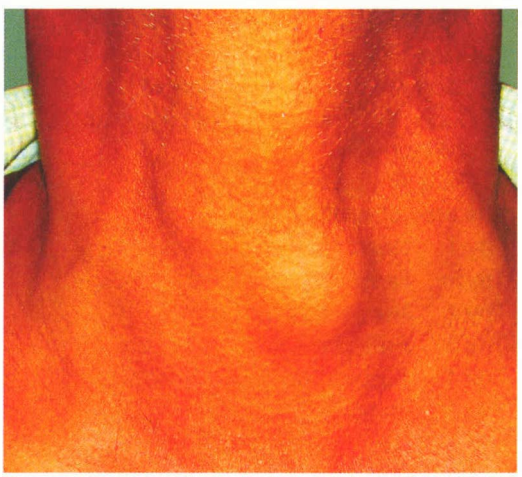


Fig. 19.65: A 45-year-old male presented with solitary nodule. Hormone levels were elevated. He underwent hemithyroidectomy—solitary toxic nodule

b. STG level above 50 µg/L—suggests residual (or) recurrent tumour
c. STG level goes above 100 µg/L suggests pulmonary (or skeletal metastasis)
d. Increased in hyperthyroidism, thyroiditis and tumour
e. Serum calcitonin—useful for screening medullary carcinoma when a patient has history of thyroid cancer and presents with a solitary nodule

2. 24 hours collection of urine for catecholamine to exclude phaeochromocytoma (MEN II syndrome) because of association with medullary carcinoma.
3. **Ultrasonography (USG):** It is noninvasive, cheap and it has become the investigation of choice in solitary nodule (Key Box. 19.36).

Following patterns are described

- Totally sonolucent unilocular lesion
- Sonolucent cyst with internal echoes, septae
- Nodule with homogeneous echogenicity (hyper or hypo)
- Mixed echogenicity

KEY BOX 19.36

FEATURES SUGGESTIVE OF MALIGNANCY IN USG

- Sonolucent cyst with projections of echogenic solid nubbins.
- Rich in vascularity
- Rich in microcalcification
- Can detect jugular lymphadenopathy
- Ultrasonography guided FNAC of gland and lymph node can be done.
- Cyst larger than 3–4 cm, 14% chances of malignancy.

USG has almost replaced radio-nuclide scan in the evaluation of solitary nodule.

4. **Fine needle aspiration cytology (FNAC)** (Key Box 19.37)

KEY BOX 19.37

FOLLOWING PATTERNS ARE OBSERVED

- Class I : Paucicellular aspirate rich in colloid
- Class II: Paucicellular smear rich in follicular cells.
- Class III: Frankly malignant (papillary and medullary carcinoma)
- Class IV: Indeterminate

It is also classified as

Thy 1	Nondiagnostic
Thy1c	Nondiagnostic cystic
Thy 2	Non-neoplastic
Thy 3	Follicular
Thy 4	Suspicious of malignancy
Thy 5	Malignancy

5. **CT Scan:** Expensive investigation involving exposure to ionising radiation is not done routinely. Specific indications are:
   - Doubt about the origin of the swelling
   - Large lesion with local infiltration
   - Intrathoracic extension
6. **Isotope scan** (not routinely done nowdays)—Mos commonly used isotopes—$^{99m}T_c$. ^{131}I should not be usec for evaluation of nodule. ^{123}T will not detect nodules o 1 cm size. However, it is indicated in a 'toxic nodule' whicl is 'hot'. Rest of the gland is suppressed, so that such patien can be managed easily by radioiodine. It can demonstrate 3 different patterns as follows (Figs 19.66 and 19.67):
   - ***Hot nodule:*** The gland does not take up isotope but the nodule takes it up, which is a feature of **autonomous solitary toxic nodule**. Here, the normal thyroid tissue is suppressed. Localisation of over activity in a nodule with suppression of remainder of the gland is characteristic of solitary toxic nodule. In toxic MNG, multiple areas show overactivity.
   - ***Warm nodule:*** The entire gland takes up isotope. This is typical of Graves' disease (primary thyrotoxicosis) wherein each cell is active and equally stimulated.
   - ***Cold nodule*** is a nodule which does not take up isotope. It should be remembered that only 10% of the cold nodules are malignant. Keeping this in mind, surgeons do not investigate MNG with isotope scan. The assessment of malignancy is done clinically (Key Box 19.38).
7. **T3, T4:** In toxic goitre, T4 level should be assessed. T3 is useful where T4 level is borderline (T3 toxicosis)

KEY BOX 19.38

COLD NODULE—DIFFERENTIAL DIAGNOSIS

1. Haemorrhage
2. Carcinoma
3. Thyroiditis
4. Thyroid cyst

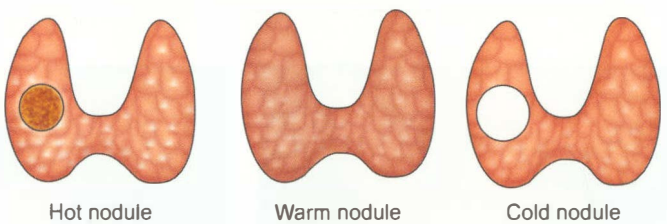


Fig. 19.66: Radioisotope scan findings

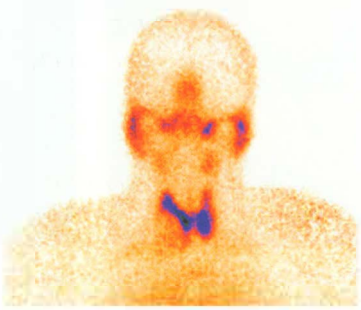


Fig. 19.67: Solitary thyroid nodule—cold in nature

Treatment

The final decision about treatment of solitary nodule depends upon the clinical findings, sonography findings and FNA cytology results.

• Please read the clinical notes.

CLINICAL NOTES

CASE OF THYROID SWELLING

A 31-year-old female with solitary nodule (2 cm) underwent FNAC—reported as adenoma. She underwent hemithyroidectomy in a peripheral hospital which was reported as papillary carcinoma thyroid (PCT). It was a histological surprise. How to proceed now?

- *At our hospital ultrasound neck was done again which could not reveal any abnormality. She underwent completion thyroidectomy (of the opposite lobe) after 4 weeks (re-exploration is done within 7 days or after 4 weeks once tissue oedema subsides completely). Thus, the patient underwent total thyroidectomy. HPR of opposite lobe did not show papillary carcinoma. No lymph node dissection was done. The patient was not given T4 for 6 weeks.*
- *After 6 weeks, ^{131}I scan was done giving 1 mCi, residual thyroid tissue was found in neck. It was ablated with 100 mCi of ^{131}I.*
- *Even though the aim of surgery is to remove the entire thyroid gland (total thyroidectomy), some parts of gland might have been left behind by the surgeon, specially in the region of ligament of Berry. Hence, a routine post-operative radionuclide scan is advisable.*
- *After ablation, she was put on T4 0.3 mg/day to be taken in the morning on an empty stomach.*
- *She came after 6 months with pain in the neck for which ultrasound was done. It revealed suspicious residual thyroid tissue.*

What to do now?

- *Serum thyroglobulin was estimated. It was < 0.2 (almost undetectable limits; Normal 0.2–55). So, the suspicious lesion could have been some fibrous tissue due to surgery. As thyroglobulin was normal, we did not investigate her further.*
- *Now it is 3 years since completion of thyroidectomy and the patient has no disease but is on T4 0.3 mg/day.*

What are the annual tests to be done?

- *Serum TSH, calcium, thyroglobulin, ultrasound, Thyroglobulin antibody and if necessary, nuclear scan of neck.*

COMMENTS

I have heard in a few surgical meetings, the following statement made by a surgeon, 'I have done total thyroidectomy, how can residual thyroid tissue be present in the neck?' Remember, it is always possible that some thyroid tissue is left behind especially in the region of ligament of Berry, tracheoesophageal groove and posteromedially.

It is largely classified into two groups: One, where FNAC results are available and the other, where they are not available.

I. FNAC results available

Examples	Treatment
• Papillary carcinoma	: Total thyroidectomy with central neck dissection
• Medullary carcinoma	: Total thyroidectomy with the modified radical neck dissection
• Adenoma	: Lobectomy (hemithyroidectomy)
• Toxic nodule in young	: Lobectomy
• Toxic nodule in elderly	: Lobectomy or ^{131}I ablation
• Simple cyst	: If it resolves after aspiration, wait and watch. If it recurs, do a lobectomy
• Colloid goitre	: Observe, L-thyroxine 0.1 to 0.2 mg/day.

Central neck dissection

- Dissection should include ipsilateral tracheo-oesophageal groove
- Pretracheal area: Along recurrent laryngeal nerve and inferior thyroid vein
- Anterior mediastinum

II. FNAC results not available or tissue inadequate: In such cases, it can be repeated and sonography guided FNAC should be done. If this is also inconclusive, following management plan can be offered:

- Follicular cells seen: Total thyroidectomy. Even if the final report is benign it is acceptable (**this is described as overtreating**).
- If the lesion is cystic and the patient is a female, it is less likely to be malignant and can be observed.
- If the lesion is **solid and the patient is a male**, it needs to be explored (Key Boxes 19.39, 19.40 and Fig. 19.68).

KEY BOX 19.39

'MOST COMMON' FOR SOLITARY NODULE THYROID

- Most common surgical disease of thyroid gland is solitary nodule
- Most of the thyroid diseases begin as solitary nodule.
- Most common investigation of choice for solitary nodule is ultrasonography.
- Most common cause of a solitary nodule is multinodular goitre.
- Most common cause for solid solitary nodule in a male is malignancy.
- Most commonly used investigation of choice for tissue diagnosis is FNAC.

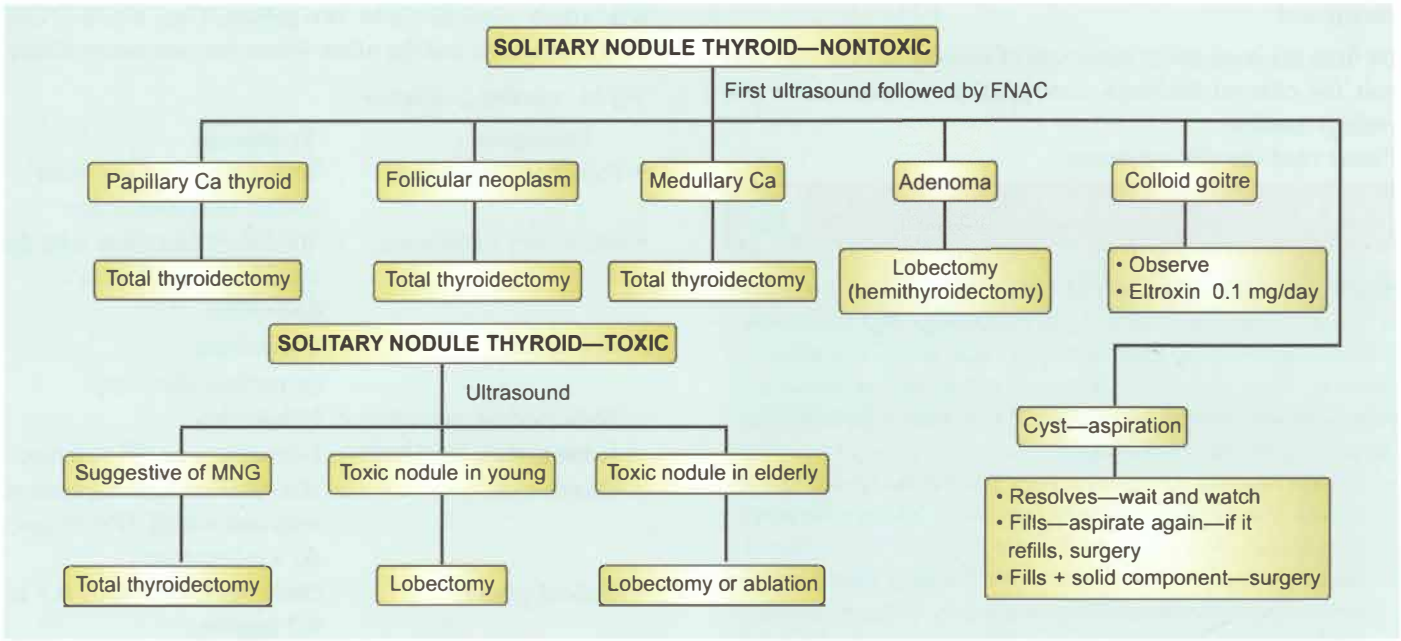


Fig. 19.68: Algorithm of treatment plan in solitary thyroid nodule

KEY BOX 19.40

WHAT SHOULD NOT BE DONE IN A NODULE

- Nodule should never be enucleated
- Male solitary solid nodule should never be left alone for observation
- Suppression therapy should never be attempted for a solitary nodule without a definitive diagnosis.
- Radio-ablation/suppression therapy should never be given to a nodule causing compression on the trachea.
- FNAC should not generally be done to a nodule, if it is a post-irradiated gland or there is familial thyroid cancer.

THYROID INCIDENTALOMA

- As the name suggests these are incidentally detected during head and neck ultrasonography done for some other problems.
- In such situations, the management is similar to that of a solitary nodule.
- Any lesion more than 1.5 cm and lesion which is solid should be subjected to sonography-guided FNA.
- Depending on the results of the FNA, treatment is recommended.
- If it is < 1.5 cm, just follow up with annual sonogram.

In western countries with increasing obesity, and short neck, it is not surprising to see many such cases of incidentaloma detected by ultrasound.

THYROIDITIS

- Bacterial infection of thyroid is very rare because of its rich blood supply.
- Thyroiditis is broadly classified into granulomatous autoimmune and Riedel's thyroiditis (Table 19.9).

GRANULOMATOUS THYROIDITIS

- It is also called subacute thyroiditis or de Quervain's disease
- This occurs due to viral infection. It usually follows sore throat (mumps virus has been incriminated in a few cases)
- Patients present with fever, body ache and painful enlargement of thyroid gland. The gland is enlarged, tender to touch, soft to firm and a few symptoms of hyperthyroidism occur initially.
- ESR is increased.

Treatment

Majority of the patients respond to conservative treatment in the form of analgesics and a short course of prednisolone. There are no permanent sequelae of this condition.

AUTOIMMUNE THYROIDITIS

- Hashimoto's thyroiditis is the main component of thyroiditis.
- Autoimmune aetiology is characterised by extensive lymphocytic infiltration resulting in destruction of thyroid follicles with variable degree of fibrosis.
- Females in perimenopausal group (40–50 years) are commonly affected. Initially, symptoms of mild hyperthyroidism (hashitoxicosis) may be present. Later, extensive intrathyroidal fibrosis results in permanent hypothyroidism.
- The thyroid follicles are destroyed by significant fibrosis. The deep eosinophilic-staining thyroid follicular cell, Askanazy cell, is characteristic of Hashimoto's thyroiditis.

Table 19.9 Comparison of three forms of thyroiditis

	Granulomatous	Hashimoto's	Riedel's
1. Aetiology	Virus	Autoimmune	Collagen disorder
2. Age group	Young	Woman at menopause	Old age
3. Pathology	Inflammatory cells	Lymphocytes, fibrosis	Extensive fibrous tissue
4. Clinical	Painful, tender, smooth, sudden goitre	Irregular or nodular, firm nontender	Hard, irregular, fixed, nontender
5. Toxicity	Initial toxicity, later normal	Initial toxicity, later hypothyroidism	Hypothyroidism
6. Laboratory tests	ESR is increased	Antithyroid antibodies ↑	No biochemical test
7. Treatment	Symptomatic	Thyroxine, surgery	Thyroxine, surgery
8. Differential diagnosis	Acute bacterial thyroiditis	Multinodular goitre	Anaplastic carcinoma

- The gland can be firm to hard and sometimes rubbery in consistency, smooth or irregular and can involve a lobe or the entire gland.
- In many cases, thyroid antibodies are raised, suggesting an autoimmune disorder.

Treatment

- Thyroxine 0.2 mg/day is given as a supplementary dose.
- If there is compression on the trachea, isthmusectomy is done to relieve compression.
- If the goitre is big and causing discomfort, subtotal thyroidectomy can also be done.

Complications of Hashimoto's thyroiditis

- Permanent hypothyroidism
- Papillary carcinoma of the thyroid
- Lymphoma

RIEDEL'S THYROIDITIS

- This is a very, very rare cause of a goitre which is supposed to be a collagen disorder.
- It can be associated with **mediastinal fibrosis, retroperitoneal fibrosis and sclerosing cholangitis**.
- In this condition, there is intrathyroidal fibrosis but extrathyroidal fibrosis is more.
- Involvement of trachea, oesophagus, internal jugular vein, carotid artery, etc. result in dysphagia and dyspnoea.
- As a result of fibrosis, all the thyroid follicles are replaced by fibrous tissue.
- By the time patients present to the hospital, it is an advanced stage and excision is very difficult.

Treatment

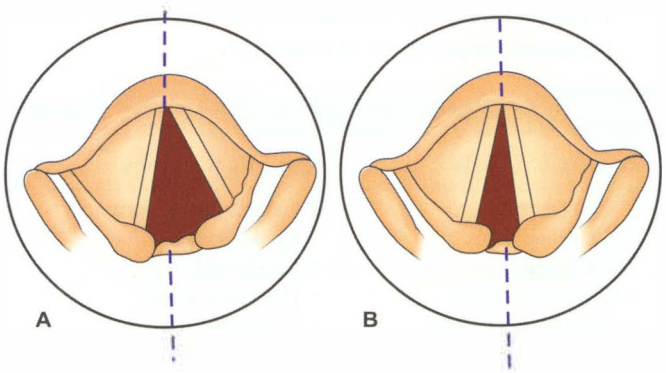
- Treatment with thyroxine may be necessary to treat hypothyroidism.
- In selected difficult cases, isthmusectomy can be tried to relieve compression on the trachea. Three types of thyroiditis are compared in Table 19.9.

COMPLICATIONS OF THYROIDECTOMY

1. **Haemorrhage** can be a primary haemorrhage which occurs during surgery.
   - Reactionary haemorrhage is more dangerous and occurs within 6–8 hours after surgery. This is due to slipping of ligature because of straining, coughing, hypertension, etc.
   - It is a tension haematoma which develops deep to deep fascia, compressing the larynx. Re-exploration of neck under GA, control of bleeding points and evacuation of haematoma should be done immediately.
   - Without evacuating haematoma, an attempt to intubate the patient may result in cardiac arrest (as such patient will be struggling).
2. **Respiratory obstruction** can be due to tension haematoma resulting in compression of the larynx, collapse, tracheal cartilage softening (tracheomalacia).
   - Endotracheal intubation and a short course of steroid therapy is necessary.
3. **Laryngeal nerve paralysis** (Tables 19.10 and 19.11)

   A. **Unilateral recurrent laryngeal nerve (RLN) palsy** produces a whispering voice. The opposite vocal cord compensates. There will not be problems of aspiration or airway obstruction (Fig. 19.69A).

   B. **Bilateral recurrent laryngeal nerve palsy:** It is also known as bilateral abductor paralysis. Both the vocal cords come to be in median or paramedian position, the airway is inadequate causing dyspnoea and stridor but the voice is good (Fig. 19.69B).



Figs 19.69A and B: Vocal cord position in unilateral and bilateral recurrent laryngeal nerve paralysis

Table 19.10 Various positions of vocal cord in laryngeal nerve paralysis

Position	Inference	Distance from the mid line
1. Median	Seen in phonation	Nil
2. Paramedian	Bilateral RLN palsy	Distance: 1–2 mm
3. Intermediate (cadaveric)	Bilateral, combined paralysis of RLN and superior laryngeal nerve	Distance: 3–4 mm
4. Full abduction	Forceful inspiration	Distance: 8–9 mm

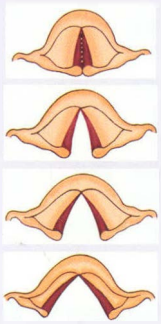


Fig. 19.70

Treatment: Tracheostomy is required as an emergency procedure. Following tracheostomy, the patient is followed up on an outpatient basis for a period of 8–9 months. This period is required for any spontaneous recovery. If no recovery occurs after this period, it requires a permanent solution. The choice is between a permanent tracheostomy with a speaking valve or a surgical procedure to lateralise the cord which can be done by endoscopic method. The former relieves stridor and preserves good voice but has the disadvantage of a tracheostomy hole in the neck. The latter relieves airway obstruction but at the expense of good voice. However, there is no tracheostomy hole in the neck.

Lateralisation of the cord

- It means to move and fix the arytenoid, vocal process and vocal cord into an abducted position.
- Endoscopic lateralisation can be done by laser cordotomy
- KTP-532 nm laser is used
- It is minimally invasive, single procedure, quick recovery.

C. Superior laryngeal nerve

- It is a branch of vagus-gives a 'motor' branch—external laryngeal nerve. It supplies cricothyroid which is adductor of the cord. Paralysis causes weak and husky voice and inability to raise the pitch of voice (This is of particular importance in singers).

Table 19.11 Causes of recurrent laryngeal nerve paralysis

Left RLN lesions	Right RLN lesions
• Ca bronchus • Ca oesophagus • Ca thyroid • Operative injury • Peripheral neuritis • Aortic aneurysm • Mediastinal mass • Left atrial enlargement	• Ca apex of the lung • Ca oesophagus • Ca thyroid • Operative injury • Peripheral neuritis • Subclavian aneurysm

- Large internal laryngeal nerve supplies whole supraglottic larynx—sensory innervation to the epiglottis. pyriform sinus and larynx as far down as vocal folds.

D. Combined (complete paralysis)

I. **Unilateral:** This results in paralysis of all muscles of larynx on one side. Vocal cord will be in cadaveric position. The healthy cord is unable to approximate the paralysed cord, thus causing glottic incompetence. This results in hoarseness of voice and aspiration of liquids through glottis. Cough is ineffective due to air wastage.

Treatment includes procedures to medialise the cord and speech therapy.

II. **Bilateral:** This is an uncommon condition. This results in paralysis of all the intrinsic muscles of the larynx. Both vocal cords assume cadaveric position. There is also a total anaesthesia of larynx.

PEARLS OF WISDOM

It is better to identify RLN in all cases of thyroidectomy—specially total thyroidectomy and lobectomy.

4. **Permanent hypothyroidism** can develop slowly after thyroid surgery especially after subtotal thyroidectomy for Graves' disease. It takes 2–3 years for manifestation of hypothyroidism to become apparent.

5. **Permanent hypoparathyroidism** is managed with calcium tablets or with 1, 25-dihydroxy cholecalciferol.

6. **Thyrotoxic crisis (storm)**

- Thyrotoxic storm occurs in patients with primary thyrotoxicosis who are improperly treated or prepared for surgery. At surgery, due to handling of the gland, sudden release of thyroxine into the systemic circulation results in thyrotoxic crisis (Key Box 19.41).

KEY BOX 19.41

THYROTOXIC STORM PREVENTION

- Euthyroid before surgery
- β-blockers, carbimazole
- Lugol's iodine
- Good anaesthesia
- Perfect haemostasis
- Gentle manipulation

- Hyperpyrexia—Above 105°F, severe sweating, gross dehydration, hypovolaemic shock and tachycardia are the diagnostic features.
- It is treated by following measures:
  - ICU admission and resuscitation: O_2, correction of dehydration by rapid IV fluids.
  - Cold tepid sponging, to control the temperature.
  - IV and oral propranolol 2–4 mg and as necessary.
  - Hydrocortisone 100 mg, thrice daily.
  - Carbimazole or propyl thiouracil
  - In spite of the above treatment, mortality is high.

PEARLS OF WISDOM

Prevention is better than 'cure'. With adequate preparation, thyrotoxic storm can be prevented.

7. **Wound infection:** It is not common to get wound infection after thyroid surgery. However, antibiotics are started if there is evidence of local erythema, tenderness and if the patient has fever.
8. **Scar hypertrophy and keloid**
9. **Stitch granuloma:** May occur with/without sinus formation and is seen after the use of nonabsorbable suture material. Absorbable ligatures and sutures (VICRYL) can be used throughout thyroid surgery except for skin closure where silk is still appropriate.

MISCELLANEOUS

LINGUAL THYROID

- Occasionally, a patient presents with a small swelling in the middle of the tongue at the junction of anterior 2/3 and posterior 1/3 of the tongue.
- It could be lingual thyroid—an aberrant thyroid tissue found in the region of foramen caecum on the tongue.
- Foramen caecum represents the junction of epithelial floor of the mouth with proximal portion of thyroglossal duct.
- Even though lingual thyroid is rare, it can give rise to significant complications (Fig. 19.71).

Fig. 19.71: Four centimetre lingual thyroid in the classical location causing dysphagia—successfully removed with LASER

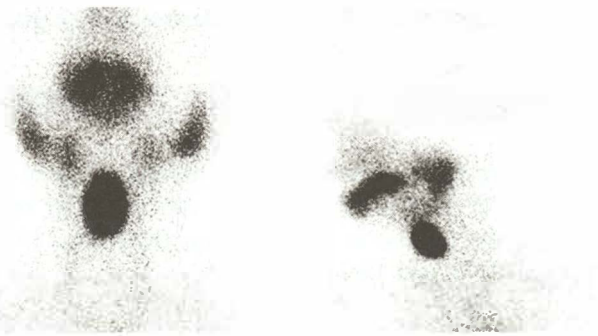


Fig. 19.72: ^{99m}Tc (Technetium) scan showing lingual thyroid

Clinical features

- Common in females (3:1)
- Swelling in the tongue in the classical location, firm in consistency, can be irregular. Impairment of speech, haemorrhage, dysphagia, airway obstruction may be the presenting features.
- Majority of patients have hypothyroidism (70%) and 10% are cretins.

Diagnosis

- Usually made on clinical grounds.
- Thyroid scan-ultrasound of neck is done to find out whether thyroid tissue is present or not (often it is absent).
- ^{99m}Tc (technetium) scan will detect ectopic thyroid tissue (Fig. 19.72).

Differential diagnosis

1. **Ectopic salivary gland tumour.** Tongue is the commonest site. To begin with mucosa is normal starts as a submucosal swelling.
2. **Gumma:** Rare nowadays. It is a midline, indurated, firm, smooth swelling, painless.

3. **Lymphangioma and haemangioma:** These are present since birth, more diffuse, fluctuant cystic swellings. Haemangioma is compressible and lymphangioma is transilluminant.

Treatment

PEARLS OF WISDOM

Lingual thyroid may be the only thyroid tissue present in a patient. Hence, a thyroid scan is done to confirm the presence of normal thyroid tissue.

- Small dose of thyroxine may decrease the size of the swelling (similar to a puberty goitre) as a treatment of hypothyroidism or to suppress TSH.
- Large swelling with significant symptoms needs to be excised. Laser excision is better.
- Radioactive iodine to suppress/destroy can be given.

ECTOPIC THYROID

- It occurs due to failure of median thyroid anlage to descend normally. Hence thyroid tissue can be found anywhere from the tongue to mediastinum (Key Box 19.42 and Figs 19.73 and 19.74).

KEY BOX 19.42

ECTOPIC THYROID TISSUE

- Lingual thyroid
- Thyroglossal ectopic thyroid in the upper part of the neck
- Adjacent to aortic arch, aortopulmonary window, within upper pericardium, etc.
- Struma ovarii—malignant ovarian teratoma containing thyroid tissue.

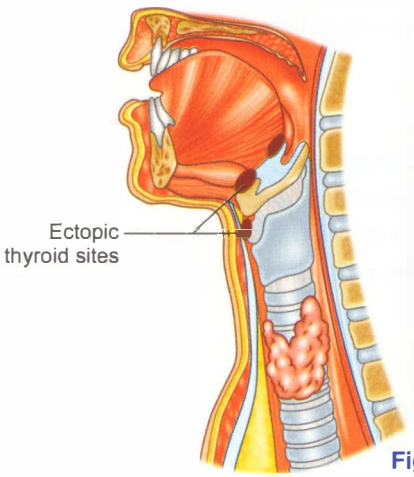


Fig. 19.73: Ectopic thyroid sites

Fig. 19.74: Ectopic thyroid tissue in the submandibular region turned out to be papillary carcinoma thyroid

- Ectopic thyroid tissue can have the diseases similar t normal thyroid tissue, such as solitary nodule, multi nodular goitre, malignancies, etc.
- Diagnosis is by ultrasound, CT scan. FNAC to confirm the diagnosis.
- Treatment depends upon the pathology.

INTERESTING 'MOST COMMON' FOR THYROID GLAND

- Most common surgical disease of the thyroid gland is solitary thyroid nodule.
- Most common site of thyroid nodule is at the junction of isthmus and lobe.
- Most commonly used drug for thyrotoxicosis is carbimazole.
- Most commonly used noninvasive investigation for thyroid diseases is ultrasonogram.
- Most commonly done surgery for well-differentiated carcinoma thyroid is total thyroidectomy.
- Most common thyroid cancer in children is papillary carcinoma thyroid.
- Most common thyroid cancer following exposure to external radiation is papillary carcinoma thyroid.
- Most common noninvasive investigation in the evaluation of solitary nodule is USG.

MOST DIAGNOSTIC FEATURES IN THYROID GLAND

- Most diagnostic feature of medullary carcinoma thyroid is **amyloid stroma**
- Most diagnostic feature of papillary carcinoma thyroid is presence of **psammoma bodies**
- Most diagnostic feature of Hashimoto's thyroiditis is **Askanazy cell**
- Most diagnostic investigation for lingual thyroid is **^{99m}Tc (technetium) scan**
- Most diagnostic feature of anaplastic carcinoma is **early fixity**
- Most diagnostic feature of Graves' disease is exophthalmos

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been edited.
- Relevant microscopic pictures (histopathology) have been included.
- New photographs, key boxes and flowcharts have been added.
- Solitary thyroid nodule is discussed in more detail.

MULTIPLE CHOICE QUESTIONS

1. Superior thyroid artery is a branch of:
A. Common carotid artery
B. Internal carotid artery
C. External carotid artery
D. Thyrocervical trunk

2. Following are true for T3 hormone *except*:
A. It is more important physiologic hormone
B. It can be produced by peripheral conversion of T4
C. It is more quickly acting
D. It is given once a day for suppressive dose

3. Following are true for primary thyrotoxicosis *except*:
A. 8 times more common in women than men
B. Gland is vascular and smooth
C. Cardiac failure is common
D. Proptosis is a feature

4. Replacement dose of thyroxin – T4 is:
A. 0.1 mg
B. 0.2 mg
C. 0.3 mg
D. 0.01 mg

5. Papillary carcinoma thyroid spreads to which lymph nodes?
A. Submental
B. Submandibular lymph nodes
C. Scalene nodes
D. Jugular chain of lymph nodes

6. Following is not the common sign of Graves' disease:
A. Proptosis
B. Pretibial myxoedema
C. Proximal myopathy
D. Cardiac failure

7. The widely used first investigation of choice in solitary nodule thyroid is:
A. X-ray neck
B. CT scan
C. MRI
D. Ultrasound

8. Which one of the following is not the treatment for Graves' disease?
A. Carbimazole
B. Propranalol
C. Massive doses of steroids in ophthalmopathy
D. Digoxin

9. Following are true for treatment options for toxic goitre *except*:
A. Thyroidectomy
B. Antithyroid drugs
C. Radioiodine treatment
D. Radiotherapy

10. Which one of the following is the treatment of choice for toxic goitre in a child?
A. Radioiodine
B. Thyroidectomy
C. Antithyroid drugs
D. Radiotherapy

11. Which is the most effective drug for thyrotoxic storm?
A. Steroids
B. Noradrenaline
C. IV propranalol
D. Carbimazole

12. Which one of these operative steps is not done routinely in thyroidectomy?
A. Ligation of middle thyroid vein
B. Ligation of superior thyroid artery
C. Ligation of inferior thyroid artery
D. Ligation of inferior thyroid veins

13. Few landmarks for identification of recurrent laryngeal nerve include following *except*:
A. Parallel to trachea-oesophageal groove on the left side
B. Below the inferior thyroid artery on the right side
C. Between branches of the inferior thyroid artery
D. Between the branches of inferior thyroid veins

14. Following are true for external branch of superior laryngeal nerve *except*:
A. It is close to superior thyroid artery
B. Paralysis of this nerve leads to loss of tension in the vocal cord
C. Paralysis of this nerve diminishes power and range in the voice
D. It supplies adductors of the vocal cord

15. Following are true for papillary carcinoma thyroid *except*:
A. Childhood irradiation is an important cause
B. Orphan Annie-eyed nuclei is characteristic
C. Multiple foci are common in the same lobe
D. It spreads predominantly by blood spread

16. Single most important aetiological factor in papillary carcinoma is:
A. Endemic goitre
B. Childhood irradiation
C. Autoimmune thyroiditis
D. Multiple endocrine neoplasia syndromes

17. Following are the important steps of surgery for well-differentiated thyroid cancers of more than 2 cm in size *except*:
A. Lobectomy
B. Total thyroidectomy
C. Selective neck dissection
D. Removal of all macroscopic disease

18. Following are true after total thyroidectomy before radioiodine scan:
A. If T_3 is given, only one week of stopping of hormone is enough
B. Otherwise minimum 4 weeks stoppage of T4 is required
C. Alternative to thyroxine withdrawal is to administer synthetic TSH over a 48 hours
D. TSH administration is not at all effective

19. Following are true about thyroglobulin *except*:
A. Normal levels are not detectable
B. Increasing levels indicate metastasis in papillary carcinoma thyroid
C. Increasing thyroglobulin prompts surgeon to get ultrasound of the neck
D. Useful in medullary carcinoma thyroid

20. Following are true for medullary carcinoma thyroid *except*:
A. Calcitonin is the tumour marker
B. It arises from parafollicular cells
C. It has amyloid stroma
D. It can be treated by lobectomy

21. Following is an indication for core needle biopsy o thyroid swellings:
A. Lymphoma
B. Papillary carcinoma thyroid
C. Follicular carcinoma thyroid
D. Medullary carcinoma thyroid

22. Following are pathological features of papillar carcinoma *except*:
A. Microcarcinoma
B. Orphan Annie-eyed nucleai
C. Tall cell cancer
D. Hürthle cell cancer

23. Following are true for medullary carcinoma thyroi *except*:
A. It arises from parafollicular cells
B. Cells are like carcinoid tumour
C. Does not spread by lymphatics
D. Tumours are not TSH dependent

24. Follicular carcinoma thyroid is diagnosed by:
A. Orphan Annie-eyed nuclei
B. Askanazy cell
C. Angioinvasion and capsular invasion
D. Amyloid stroma

ANSWERS

1 C	2 D	3 C	4 A	5 D	6 D	7 D	8 D	9 D	10 C
11 C	12 C	13 D	14 D	15 D	16 B	17 A	18 D	19 D	20 D
21 A	22 D	23 C	24 C						

20

Parathyroid and Adrenals

- Surgical anatomy
- Congenital anomaly
- Physiology—calcium and action of PTH
- Tetany
- Hyperparathyroidism
- Acute hypercalcaemic crisis
- Adrenal glands—anatomy, physiology
- Disorders of adrenal cortex
- Neuroblastoma
- Phaeochromocytoma
- Incidentaloma
- What is new?/Recent advances

PARATHYROID GLANDS

SURGICAL ANATOMY

- These are the endocrinal glands situated in the neck and secrete the hormone parathormone (PTH). Their secretion is **not dependent on pituitary gland**.
- They are 4 in number; 2 on the right and 2 on the left. They are pinkish in children, and yellow to brown in adults.
- **Superior parathyroids** are ***derived*** from endoderm of **4th branchial arch** and thus, they develop along with the thyroid gland.
- Superior parathyroids are found in relation to inferior thyroid artery in the middle of posterior aspect of thyroid gland. They are situated in the fat above inferior thyroid artery, close to cricothyroid artery and cricothyroid articulation. They are constant in position (Fig. 20.1).
- They are smaller (20-40 mg). They are **dorsal to recurrent laryngeal nerve** (RLN).
- **Inferior parathyroids** develop from endoderm of **3rd branchial arch** (with thymus) and are not constant in position. They may be seen in the lower pole, within fascial sheath of thyroid gland, low down in the neck (rarely in the mediastinum), outside the fascial sheath or even within thyroid gland. They are large (30 to 50 mg). They are **ventral to RLN** (Fig. 20.2).
- Parathyroids appear golden-yellow to light brown in adults.

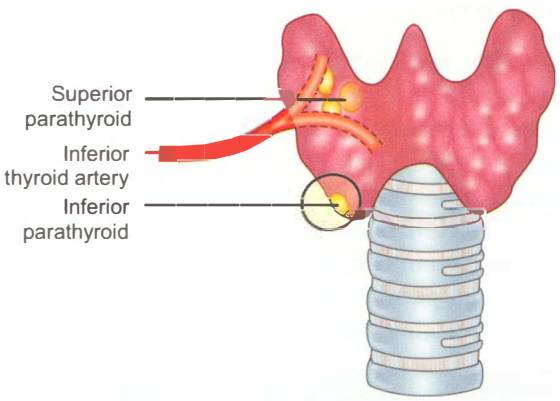


Fig. 20.1: Sites of parathyroid glands

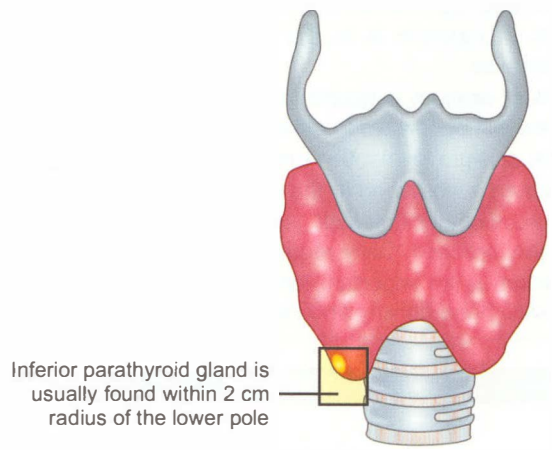


Fig. 20.2: Location of inferior parathyroid gland

Blood supply

Inferior thyroid artery supplies both the parathyroids in about 95% of cases by a leash of vessels. Ligature of both inferior thyroid arteries **may not result in hypoparathyroidism** because there is adequate collateral circulation and few branches of superior thyroid artery also supply 20% of upper glands.

Histology

Principal cells (chief cells) are the cells which secrete PTH (parathormone). Water clear cells are found in hyperplastic and neoplastic glands (rich in glycogen).

PTH

It is a peptide containing 84 amino acids. It is secreted in response to increase in calcium or decrease in magnesium. Calcitonin has 'NO ROLE' in human beings.

CONGENITAL ANOMALY AND SYNDROME

DiGeorge syndrome: Congenital absence of the parathyroid glands and thymus. Neonatal hypoparathyroidism along with absence of thymus-dependent lymphoid system.

PHYSIOLOGY—CALCIUM AND ACTION OF PTH

Action of PTH

1. Resorption and mobilisation of calcium from the bone (Key Box 20.1).
2. Increased reabsorption of calcium from kidney prompting excretion of phosphate.
3. Enhances absorption of calcium from the gut.
   - Thus, PTH increases serum calcium level.
   - Normal serum calcium level 9–11 mg/dl (total).
   - In normal persons, the PTH is balanced by calcitonin, secreted from *C-cells* of the thyroid glands.

KEY BOX 20.1

CALCIUM

- Most abundant cation in human beings
- 50% of calcium is in the ionised form, which can be measured
- 50% of calcium is bound to albumin. Hence, total calcium levels will be low when albumin levels are low
- Total calcium levels: 9–11 mg/dl
- Calcium is absorbed in the small intestines
- Sources of calcium: Milk, yoghurt and cheese, green peas, beans, oranges. Daily requirement: 1000–1300 mg/day
- Calcium is an important ion for blood coagulation, cellular activity, bone density and neuromuscular activity

TETANY

Tetany is a condition wherein there is hyperexcitability of peripheral nerves.

Causes of tetany

1. **Hypoparathyroidism**: It results from surgical removal o parathyroids. Subtotal thyroidectomy and near-tota thyroidectomy are the most common causes of tetany Hypoparathyroidism may be 'familial' variety or neonata variety. It may be temporary after thyroidectomy o permanent if all the 4 parathyroids are removed or deprivec of their blood supply. The incidence is around 1–2%.
2. **Severe respiratory alkalosis** can cause tetany as ir ***hyperventilation.***
3. Low calcium levels can occur due to dietary factors or poo absorption from the gut or ***acute pancreatitis, chronic rena failure,*** etc.
4. **Osteomalacia** and **rickets** due to deficiency of ***vitamin D***.
5. **Hypokalaemic alkalosis** of pyloric stenosis.
6. Following massive transfusion (citrate overload).

Signs and symptoms (Key Box 20.2)

1. Tingling and numbness of the fingers, toes, lips (circum oral paraesthesia) and occasionally, with circumoral pallor.
2. Cramps of the hands and feet.
3. In severe hypocalcaemia, there may be ***carpopedal spasm***. **Metacarpophalangeal (MP)** joints are flexed, **inter-phalangeal joints** are extended and the thumb is adducted. This is called *thumb in palm deformity* **(obstetrician's hand)**. In the foot, extension in the ankle joints and flexion of the toes is seen.
4. **Stridor** is a dangerous complication of severe tetany due to spasm of muscles of respiration.
5. ***Latent tetany*** can be diagnosed by:
   - Tapping the facial nerve at the angle of jaw. This produces twitching of eyelids, corner of the mouth, etc. It is called **Chvostek's sign**. It indicates facial nerve hyperexcitability.
   - **Trousseau's sign:** When a blood pressure cuff applied to the arm is inflated above the systolic pressure (200 mm of Hg), the hand and feet go into spasm (obstetrician's hand, carpopedal spasm).

KEY BOX 20.2

HYPOCALCAEMIC TETANY

Trousseau's sign—carpopedal spasm
Excitability—neuromuscular
Tonic—clonic seizures—spasm, laryngeal stridor
Anxiety, confusion, depression
Numbness and tingling—circumoral
Y Chvostek's sign **Y**es (positive)
Remember as **TETANY**

6. Spasm of intraocular muscles results in blurring of vision.
7. **Convulsions**, even though rare, can occur in infants.

Diagnosis

This is established by estimating **serum calcium level** which is usually < 7 mg%.

Treatment

1. Oral calcium such as calcium lactate, calcium gluconate may relieve mild symptoms.
2. In **acute cases**, **injection calcium gluconate** 10% (10 ml) should be given *slowly intravenously* over 10 minutes to avoid cardiac arrhythmias.
3. If any precipitating cause is detected, it needs to be corrected.

HYPERPARATHYROIDISM (HPT)

Hyperparathyroidism is an uncommon disease and occurs due to an increased activity of parathyroids and manifests as hypercalcaemia.

Types of hyperparathyroidism

- **Primary hyperparathyroidism:** It refers to hyperactivity of parathyroids due to an adenoma or primary hyperplasia of parathyroid glands.
- **Secondary hyperparathyroidism:** It occurs due to persistently low levels of calcium as in chronic renal failure and malabsorption, which results in decreased levels of calcium and increased levels of hyperphosphataemia.
- **Tertiary hyperparathyroidism:** This is seen in patients who undergo dialysis and transplantation for chronic renal failure. After a few years, autonomy develops and the secondary hyperparathyroidism changes into tertiary hyperparathyroidism. They can also have problems similar to primary hyperparathyroidism.

PRIMARY HYPERPARATHYROIDISM

Aetiology

- Exact cause of primary hyperparathyroidism (PHPT) is unknown.

1. **Familial/genetic:** Can be a part of MEN syndrome - I and MEN IA or familial PHPT with jaw tumour syndrome. Both occur due to genetic mutation.

PEARLS OF WISDOM

Primary HPT is the earliest and most common manifestation of MEN I and develops in 80 to 100% of these patients by 40 years of age.

2. Low dose therapeutic ionising radiation is also one of the causes.

KEY BOX 20.3

PATHOLOGY

- Adenoma—reddish brown, only 1 gland is enlarged
- Chief cell hyperplasia—reddish brown, > 1 gland is enlarged
- Water cell hyperplasia—chocolate brown, > 1 gland is enlarged.
- Multiple gland enlargement is a feature of familial disease.

3. Lithium is associated with parathyroid hyperplasia and primary HPT. Interestingly there is no hypercalciuria and bone mineral density is not affected.
- Renal leak of calcium and declining renal function with age may also be causative factors for primary HPT.

Pathology (Key Box 20.3)

- Single chief cell adenoma is the most common cause (80%). It can be due to **diffuse hyperplasia** involving all 4 glands (5 to 10% of cases).
- Very rarely, it can be due to carcinoma arising in the parathyroid glands (1%).
- Adenoma can be a part of **multiple endocrine neoplasia** (MEN) syndrome.

Clinical features

- Common in *females:* Female to male ratio is 4:1.
- Age: 30–60 years, the most common age group is 5th decade—middle aged
- Incidence is 1:1000 patients.
- **The most common** presentation is **asymptomatic hypercalcaemia** in about 50% of the patients and **renal stones** in 25% of the patients. The clinical features are as follows:

1. Bone disease—painful bones

- Due to increasing levels of PTH, extensive *skeletal* decalcification occurs. This results in bony pains, *pathological fractures* due to brittle bones, *subperiosteal* erosions, cysts in the phalanges, mandible, skull, etc. They are called **pseudotumours**. The changes are similar to that seen in ***osteitis fibrosa cystica*** (von Recklinghausen's disease). Thus, osteopaenia, osteoporosis and osteitis fibrosa cystica are found in 15% of PHPT.

2. Renal disease—renal stones

- Increased calcium levels result from increased calcium absorption from the kidneys. Hence, patients are prone to develop **renal stone and nephrocalcinosis** (calcification of kidney), hypertension.
- Calcium also **increases the tone of the vessels** which adds to the hypertension. Primary hyperparathyroidism is the cause of stones in 1–3% of all patients with kidney stones and in 10% of those who have recurrence of stones.
- **Calculi** are specially composed of **calcium phosphate or oxalate.**

3. Abdominal groans

- Calcium stimulates **gastrin** which is a powerful stimulator of acid. This may result in pain abdomen due to peptic ulcer. The patient can present with dyspeptic symptoms.
- Calcium can cause **pancreatitis**, resulting in pain radiating to the back.
- Metastatic calcification is also a feature.
- Increased incidence of gallstones has been reported as a consequence of increase in secretion of calcium in the bile-calcium bilirubinate stone.

4. Psychiatric moans

- Hypercalcaemia can result in depression, fatigue, anxiety, psychosis and even coma.
- These patients, more often women, mostly middle age, having bony pains, backaches and behavioural abnormalities are thought to have a psychiatric illness. They are referred to mental institutions, orthopaedic department, gynaecology department and are shunted from doctor to doctor.

5. Fatigue overtones

Many patients present with weakness and fatigue.

PEARLS OF WISDOM

PHPT is disease of stones, painful bones, abdominal groans, psychic moans and fatigue overtones.

Acute presentation of hypercalcaemia

- Abdominal pain
- Oliguria
- Dehydration
- Vomiting
- Coma

Symptoms

• Asymptomatic	50–60% of cases
• Renal stones	25–30%
• Bone disease	8–10%
• Joint pains	3–5%
• Abdominal pain (peptic ulcer)	3–5%
• Hypertension	3–5%

Other features (Key Box 20.4)

- Corneal calcification/band keratopathy (inside iris) may be seen in the eye on **slit-lamp examination**.

KEY BOX 20.4

PRIMARY HYPERPARATHYROIDISM AND CALCINOSIS

- Nephrocalcinosis and renal stones
- Cholelithiasis
- Chondrocalcinosis
- Calcification at ectopic sites such as blood vessels, cardiac valves, skin, etc.—Metastatic calcification

* Soft tissue calcification and tumoural calcinosis occur in secondary hyperparathyroidism.

- **Proximal myopathy,** muscle wasting is also seen
- Interestingly, clinical examination of the neck *may no reveal* any parathyroid enlargement. Hence, the diagnosi should be suspected by the various symptoms. High inde of suspicion is necessary in arriving at a proper diagnosis

PEARLS OF WISDOM

When a patient develops multiple urinary calculi, reduced bone density, high serum calcium, symptomatic hypercalciuria and pathological fractures, suspect primary HPT and request for PTH assay.

KEY BOX 20.5

CAUSES OF HYPERCALCAEMIA

• Endocrinal	Primary hyperparathyroidism Thyrotoxicosis Phaeochromocytoma
• Malignancy	Multiple bone secondaries[1]—(Ca breast, prostate, kidney, follicular carcinoma thyroid) Multiple myeloma, oat cell carcinoma[2]
• Renal failure	Secondary and tertiary hyperparathyroidism
• Nutritional	Vitamin D intoxication
• Chronic disease	Tuberculosis sarcoidosis
• Skeletal	Paraplegia/Quadriplegia patients also are immobilised—hypercalciuric hypercalcaemia

[1]The common causes are carcinoma breast, prostate, kidney, bronchus, follicular carcinoma of the thyroid.

[2]It produces PTH-like polypeptide (pseudohyperparathyroidism).

Investigations

It can be classified as follows:

I. To prove hyperparathyroidism

- Serum calcium, phosphate, albumin
- **Serum PTH assay**
- Alkaline phosphatase
- X-ray of bones.

II. To localise parathyroid glands

- Ultrasound of neck
- Thallium and technetium subtraction scan
- Selective venous sampling with PTH assay. This is most reliable but more difficult.
- Sestamibi scanning.

I. To prove hyperparathyroidism

1. Serum **calcium** levels are always raised above normal limits (9–11 mg%). There are many causes of hypercalcaemia which are depicted in Key Box 20.5. Hence, estimation of serum calcium alone will not give the diagnosis.

2. **Albumin is the main calcium binding protein** in the plasma. Hence, it should also be measured.
3. Serum **PTH** level which is estimated by immunoassay is the *diagnostic investigation.* It is called **tumour marker** for hyperparathyroidism. Estimation of PTH is difficult, costly and needs sophisticated setup.
4. Serum **phosphorus** levels are decreased.
5. **Alkaline phosphatase** is increased when bones are involved.
6. **X-ray of the hand** may reveal decalcification cysts in the phalanges, telescoping of finger tips, etc. **X-ray of the skull** may reveal subperiosteal erosions, hazy outline of the skull **and pepper pot** appearance (Key Box 20.6).

KEY BOX 20.6

INTERESTING RADIOLOGICAL CHANGES

• Osteopaenia	• Bone density loss in cortical bone—radius
• Bone cysts and brown tumours	• Aggregation of osteoclasts (osteoclastoma)
• Rugger Jersey spine	• Generalised loss of bone density
• Pepper pot skull	• Demineralised bone—mottled appearance
• Subperiosteal resorption **(pathognomonic)**	• Seen in radial aspect of middle phalanges of 2nd and 3rd fingers, bone cysts and tufting of the distal phalanges

II. To localise parathyroid glands (Figs 20.3 to 20.6)

1. **High frequency ultrasound** of the neck can be very accurate in the hands of an experienced sonologist. It can also detect renal disease, pancreatic disease, etc. It cannot scan behind sternum and cannot pick up lesions less than 0.5 cm. Specially useful in detecting intrathyroidal parathyroid adenoma (sensitivity is about 75%).
2. **Thallium-technetium isotope scan:** First the thyroid is outlined with ^{99m}Tc and then isotope ^{201}TlCl (thallium chloride) is administered. This is taken up by both the thyroid and parathyroid. By computer subtraction of the two and enlargement of images, the parathyroid appears as a hot spot.
3. **Technetium-99m (^{99m}Tc)—labelled sestamibi (MIBI) scanning:** MIBI (methyl-isobutyl-isonitrile radionuclide—sestamibi is concentrated in tissues rich in mitochondria
   - Heart
   - Salivary glands
   - Thyroid glands
   - Parathyroid glands

   This test has proved to be superior to thallium and technetium subtraction scanning.
   - Sestamibi is a protein labelled with technetium 99m that localises diseased gland—most widely used and is an accurate modality.
   - It is **very sensitive to identify adenomas** (90%) than hyperplasia.
   - However, it is very expensive. Hence, it can be used in **'Re-exploration of neck'** for parathyroidectomy.

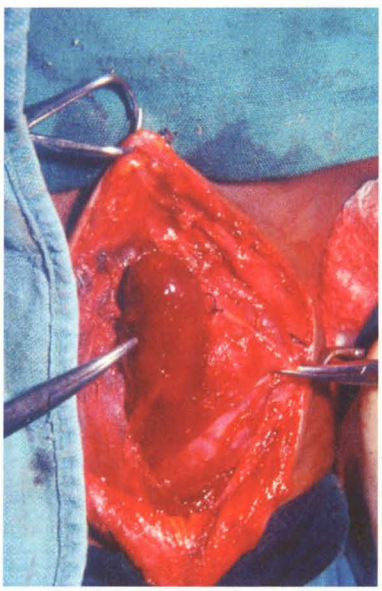


Fig. 20.3: Superior parathyroid found near the upper pole of the thyroid gland

Fig. 20.4: Proved to be adenoma—rest of the glands not enlarged

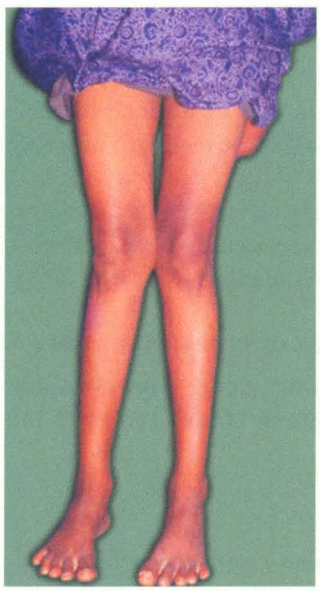


Fig. 20.5: Parathyroid adenoma with genu valgus deformity

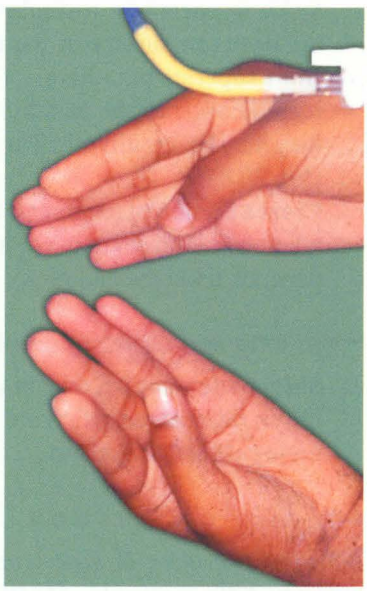


Fig. 20.6: Severe tetany following removal of the parathyroid adenoma requiring calcium infusion

(Courtesy: Prof L Ramachandra, Dr S Prasad, Dr Chethan, Department of Surgery, KMC, Manipal)

4. **SPECT: S**ingle **P**hoton **E**mission **C**omputed **T**omography when used with sestamibi is useful in evaluation of ectopic parathyroid adenomas—deep in the neck or mediastinum.

PEARLS OF WISDOM

CT scan, PET scan, MRI are not indicated prior to first time surgery.

Indications for surgery

1. Symptomatic hyperparathyroidism
2. Serum Ca 23.0
3. Renal stones
4. Hypercalciuria (> 400 mg/day)
5. Decreased creatinine clearance by 30%
6. Reduced cortical bone density

Treatment

A. Intraoperative recognition of parathyroid

1. Radioguided parathyroidectomy—it is possible if there is a preoperative MIBI scan
2. Frozen section has failure rates about 2–3%
3. Intraoperative tissue aspirate PHT—in primary HPT
4. Methylene blue is not recommended because of the risk of **toxic encephalopathy.**

B. Surgery: The surgery of the parathyroid glands needs **patience, skill** and **expertise**. The neck is explored with a collar neck incision (3–4 cm) caudal to cricoid cartilage. Parathyroid gland, when it is enlarged can be dark-brown or chocolate brown colour. Occasionally, the surgeon is lucky to encounter a single adenoma usually located on the posterior surface of the thyroid gland, when it arises from superior parathyroid (Key Box 20.7). Very often, identification of the parathyroid may be difficult because they may be intrathyroidal or within the mediastinum (Key Box 20.8).

- **Frozen section** of parathyroid glands is essential to confirm whether it is an adenoma or hyperplasia because treatment depends upon the pathology of the gland (Key Box 20.9).

Following are some examples of types of surgery

1. **Single adenoma:** Excision of the gland. **However, one other normal parathyroid gland is also removed for histopathological study**.

PEARLS OF WISDOM

Single adenoma is more common cause of PHPT in about 80% patients. Multiple adenomas are more common in older patients.

2. **Diffuse hyperplasia:** 3½ or 3¾ parathyroids are removed and **small pieces are autotransplanted into the forearm muscle tissue.** In case there is hyperactivity of this parathyroid tissue, surgical exploration becomes easy. At the same time, if this functions normally, patient will not develop hypoparathyroidism (13–14 pieces of 1 mm each in brachioradialis)
3. **Carcinoma:** All four glands should be removed along with ipsilateral thyroid lobe with modified radical neck dissection in the presence of lymph node metastasis (Key Box 20.9).

KEY BOX 20.7

IDENTIFICATION OF PARATHYROID AT SURGERY

- Bloodless field is necessary.
- Ligate middle thyroid vein. Then, retract thyroid gland medially and anteriorly.
- Identify inferior thyroid artery and recurrent laryngeal nerve.
- Parathyroids are covered by fat.
- Incise fat lobule.
- Parathyroid 'pops out'.

KEY BOX 20.8

WHEN YOU CANNOT IDENTIFY AT SURGERY

- Divide thyrothymic ligament.
- Search in tracheoesophageal groove.
- Call for intraoperative ultrasound to rule out intrathyroidal gland.
- Incise and look inside carotid sheath
- Remove thymus—send it for frozen section
- Sometimes parathyroid can be more than 4
- Do sternotomy—last step

KEY BOX 20.9

PREOPERATIVELY SUSPECT PARATHYROID CARCINOMA WHEN

- Severe symptoms
- Serum calcium levels greater than **14 mg/dl**
- Significantly elevated PTH levels
- Swelling of parathyroid gland.

Follow up

- Estimation of calcium should be done in the postoperative period to assess the functioning of the parathyroid tissue. Very often, after surgery for adenoma, there is a sudden drop in the levels of calcium because of absorption of the calcium by the bones. This is described as '**hungry bone syndrome**'. **This is seen in patients who have generalised bone disease**. In one of our patients, calcium levels dropped down to 4 mg% with severe tetany. It took 7–10 days for it to return to the normal levels. She required 24 h constant infusion of calcium.

- Absorption of calcium can be enhanced by **oral administration of 1, 25-dihydroxycholecalciferol**, which is the most active metabolite of vitamin D. Vitamin D stimulates the absorption of calcium and phosphate.

RECENT ADVANCES IN PARATHYROID SURGERY

Cervicoscopy

1. **Minimally invasive endoscopic parathyroidectomy—MIP**
   - This is possible today because of localisation by ultrasound/sestamibi combined with SPECT
   - Popularised by Gagner
   - Indicated for a single adenoma.
   - Can be done under general anaesthesia or regional—cervical block by using xylocaine (1%) with adrenaline
   - **3–4 trocars** are required
   - CO_2 insufflation is required
2. **Video-assisted parathyroidectomy—minimally invasive**
   - It is popularised by Parlo Miccoli from Italy
   - Ideal for a single adenoma **without thyromegaly**
   - No previous neck surgeries
   - It can be done without CO_2 insufflation—**gasless**
   - Bleeding, recurrent laryngeal nerve injuries are not uncommon
   - Can be done for bilateral cases
   - **No trocars**, a small 2–3 cm incision
3. **Totally endoscopic parathyroidectomy** by lateral approach by Henry, et al. It is the choice for adenoma deeper to thyroid gland. **Not for bilateral cases**. 10 mm endoscope is required. Require CO_2 insufflation.
4. **Sumary of PHPT** (Key Box 20.10)

KEY BOX 20.10

PRIMARY HYPERPARATHYROIDISM (PHPT)—INTERESTING 'MOST'

- Mostly it is sporadic.
- Mostly it is due to single adenoma.
- Most abundant cation in the body is calcium.
- Most common and earliest manifestation of MEN type I is PHPT.
- Most common presentation of PHPT is asymptomatic hypercalcaemia.
- Most widely used and accurate investigation for localising parathyroid gland is Sestamibi scan.
- Most commonly done surgery for PHPT is excision of adenoma.

CLINICAL NOTES

A 20-year-old boy attended dental outpatient department for a loose tooth. He was diagnosed to have a loose tooth and a cyst in the lower jaw as confirmed by X-ray. Patient also had genu valgus deformity with backache (Fig. 20.5). An ortho opinion was sought for. Investigations revealed high calcium levels. An ultrasound neck revealed a parathyroid adenoma of 6 cm. At exploration a single adenoma was found arising from right inferior parathyroid and was excised. Incidentally, X-ray abdomen also revealed bilateral nephrocalcinosis. This case illustration has been given to highlight the various presentations of hyperparathyroidism. High degree of suspicion is necessary for a clinician to consider the possibility of hyperparathyroidism.

ACUTE HYPERCALCAEMIC CRISIS

- It presents with severe abdominal pain and effortless vomiting. Dehydration, oliguria, renal failure follow soon.
- Untreated cases develop coma and cardiac arrest.

Causes

1. **Hyperparathyroidism**
   - Sudden increase in PTH levels occur due to spontaneous bleeding in a parathyroid tumour or rupture of a cystic parathyroid tumour. Severe dehydration also precipitates a crisis.
2. **Disseminated carcinoma** with bony metastasis (usually carcinoma of the breast).

Treatment

1. Restore fluid volume urgently.
2. The **bisphosphonates**—disodium pamidronate slow 15–60 mg single IV infusion or over 2–4 days. Maximum dose is about 90 mg. This drug stops mobilisation of calcium from the bone
3. Mithramycin.
4. Steroids (in cases of vitamin D intoxication and sarcoidosis).

- Mithramycin rapidly controlled the hypercalcemia of malignant disease in every patient. This control was temporary, and intermittent administration of the antibiotic was required. Hypercalcaemia frequently responds to hydration and moderate doses of corticoids.
- Gallium nitrate therapy is highly effective and superior to maximally approved doses of calcitonin for acute control of cancer-related hypercalcaemia.

20-YEAR-OLD BOY WITH HYPERPARATHYROIDISM: RADIOLOGICAL FINDINGS AND IMPORTANT INVESTIGATIONS (Figs 20.7 to 20.13)

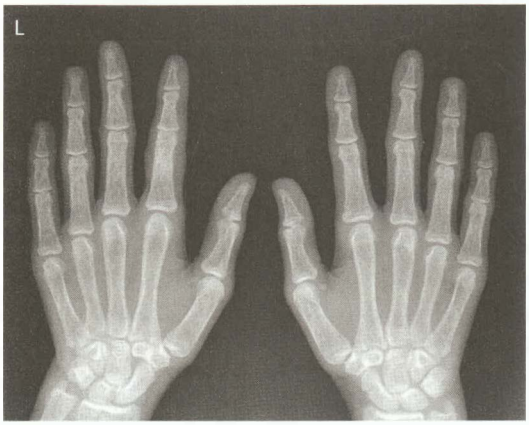


Fig. 20.7: Generalised osteopaenia and subperiosteal resorption of the radial aspect of middle phalanx—tunnelling of the cortex (double lines) is seen

Fig. 20.8: X-ray skull showing multiple lytic sclerotic lesions—pepper pot appearance

Fig. 20.9: X-ray forearm bones showing osteopaenia

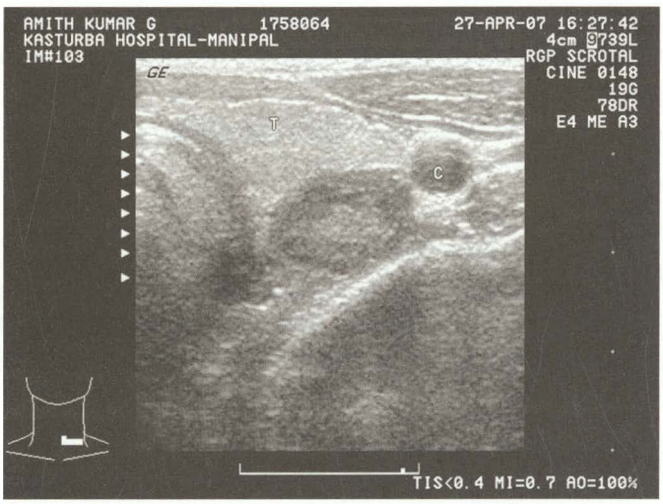


Fig. 20.10: Ultrasonography of the neck reveals heterogeneous nodule predominantly echogenic

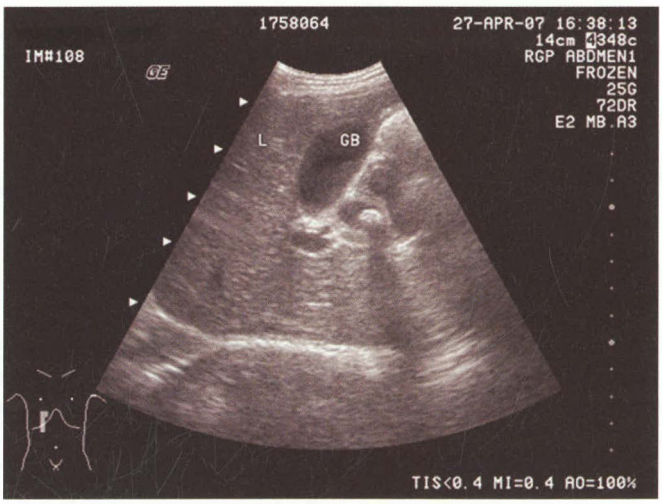


Fig. 20.11: Ultrasonography of the abdomen shows gall stone with posterior acoustic shadow

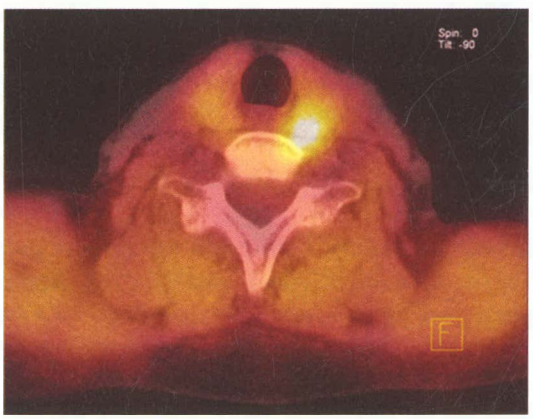


Fig. 20.12: Sestamibi scan shows bright spot—parathyroid adenoma. He also had hypercalcaemia (Key Box 20.10)

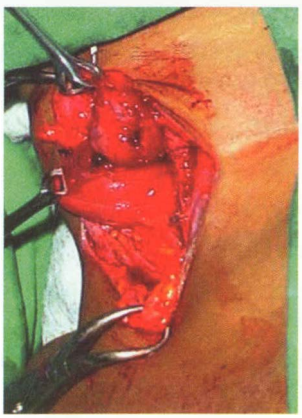


Fig. 20.13: Upper pole of thyroid gland is mobilised and 2 cm superior parathyroid adenoma is removed

ADRENAL GLANDS/SUPRARENAL GLANDS

Anatomy

- Weight of a normal gland is 4 g.
- Yellowish in colour
- Surrounded by perinephric pad of fat.
-

Right side	Left side
Pyramidal in shape	Crescent shaped
More apical	More medial to superior half of the kidney
Anterolateral to right crus	Related to spleen, tail of pancreas and left crus of diaphragm

- **Parts:** Two parts—cortex and medulla.

Blood supply

- Superior suprarenal arteries arise from **inferior phrenic** arteries.
- Middle suprarenal arteries arise from **abdominal aorta.**
- Inferior suprarenal arteries arise from renal artery.
- Suprarenal arteries branch before entering the gland so that **50 to 60 arteries** penetrate the capsule.
- **Venous drainage is through a single large suprarenal vein.** On the right side, it is short and drains into IVC. On the left side, it is longer and joined by inferior phrenic vein and empties into left renal vein.

Surgical importance

- One has to clearly identify and ligate suprarenal vein especially on the right side because of its short course. **Avulsion of right suprarenal vein** can be catastrophic.

Physiology (Table 20.1)

Cortex

- Derived from mesoderm
- Secretes corticosteroids, androgens, aldosterone.

Medulla

- Derived from neural crest cells.
- It is a mass of nervous tissue associated with sympathetic system.
- Chromaffin cells of the medulla are related to sympathetic system.
- These cells secrete catecholamines (mostly epinephrine).

Lymphatics

Drain into caval or aortic lymph nodes.

DISORDERS OF ADRENAL CORTEX

PRIMARY HYPERALDOSTERONISM (PHA)

- Triad of PHA is hypertension, hypokalaemia and hypersecretion of aldosterone (Key Box 20.11).
- PHA can be due to a **single adrenocortical adenoma** wherein it is called **Conn's syndrome.**
- It can also be due to bilateral micronodular hyperplasia
- Clinical features are headache, hypertension, hypokalaemia and weakness, commonly in women.
- Polyuria, polydypsia, nocturia are other features.
- The diagnosis is established by measuring potassium level and aldosterone to plasma renin activity ratio.
- CT scan is the most important test to locate the lesion (usually it is 1–2 cm).
- Treated by spironolactone and antihypertensive medications in bilateral hyperplasia.

KEY BOX 20.11

ALDOSTERONE

- Angiotensin II increases aldosterone secretion.
- Hyperkalaemia is another **potent stimulator of aldosterone.**
- It functions mainly to increase sodium reabsorption and potassium and hydrogen ion excretion.
- It acts at distal convoluted tubules.

Table 20.1 Histology and function

Cortex	Hormone	Functions
1. Cells of zona glomerulosa	• Aldosterone	• Regulates sodium and potassium haemostasis
2. Cells of zona fasciculata and reticularis	• Cortisol (corticosteroids)	• Gluconeogenesis • Lipolysis, fat distribution, immunological response
Medulla	• Adrenaline (epinephrine)	• Activation of CVS • Bronchodilatation
	• Noradrenaline (norepinephrine)	• Increases BP • Splanchnic vasoconstriction
	• Dopamine	• Vasodilatation in skeletal muscle • Increases heart rate

- **Laparoscopic adrenalectomy** is an effective treatment in unilateral cases.

CUSHING'S SYNDROME

- Refers to various clinical manifestations resulting from increased secretion of corticosteroids.
- When it is due to pituitary adenoma secreting large amount of ACTH—it is called Cushing's disease.
- **Ectopic ACTH producing tumours causing Cushing's syndrome are:**
  Small cell lung cancers
  Foregut carcinoid
  Medullary carcinoma thyroid
  Neuroendocrine pancreatic tumour.

Classification

- ACTH—dependent (70% pituitary or ectopic ACTH producing tumours).
- ACTH—independent: Adrenal adenoma or hyperplasia.

Clinical features (Key Box 20.12)

They are mainly due to increased corticosteroids, aldosterone and androgens. The net result is typical Cushingoid facies — with facial plethora, buffalo hump and moon face.

KEY BOX 20.12

CLINICAL FEATURES

- **C**entral obesity
- **U**nusual site obesity: Supraclavicular space, posterior neck
- **S**econdary osteoporosis: Buffalo humps
- **H**ypertension, Hyperglycaemia, Hypokalaemia, Hirsutism.
- **I**rregularity in menstruation
- **N**eurological: Depression, mania
- **G**ain in weight
- **S**kin changes: Abdominal striae, ecchymosis, acne, plethora due to thinning of subcutaneous tissues

Remember as **CUSHINGS**

Investigations

1. Morning and midnight plasma cortisol levels are increased.
2. Serum ACTH levels: If increased, it is from pituitary source or ectopic ACTH.
3. CT chest and abdomen to assess not only adrenals but also to detect ectopic sites.

Treatment

1. **Medical:** Ketoconazole or metyrapone can reduce steroid synthesis. Thus, symptoms can be controlled. They are also indicated if surgery is not possible.
2. **Trans-sphenoidal resection** of ACTH producing pituitary tumours
3. **Unilateral adenoma:** Adrenalectomy
4. **Bilateral tumours:** Bilateral adrenalectomy
   Postoperatively cortisol should be given.

PEARLS OF WISDOM

Stereotactic radiosurgery, which uses CT guidance to deliver high doses of radiotherapy to the tumour (photon or gamma knife) is being increasingly used to treat pituitary adenomas.

ADRENOCORTICAL CARCINOMA

- Incidence: One in 1,000,000 cases.
- Age: Bimodal with a first peak in childhood and second between fourth and fifth decades.
- **60%** of patients present with Cushing's syndrome.
- Diagnosis of malignancy is (> 6 cm) by size of the tumour, presence of necrosis or haemorrhage, capsular or vascular invasion.
- CT—heterogeneous tumour, irregular margins, haemorrhage, lymphadenopathy, metastasis in liver.
- MRI—angiography to exclude tumour thrombus in vena cava.
- Open radical tumour resection (R_0)—*en bloc* removal of tumour lymph nodes and involved organs is the treatment of choice.
- Adjuvant chemotherapy—**mitotane** alone (derivative of insecticide DDT) is used. It has **adrenolytic** activity.
- Etoposide, cisplatin, doxorubicin are also used.
- **Steroid hypertension** is managed by **ketoconazole**.

Adrenal insufficiency

Definition: Loss of function of adrenal cortex— is also called Addison's disease.

- Diseases associated with Addison's disease include
  1. After bilateral adrenalectomy
  2. Tuberculosis
  3. Metastases
  4. Haemorrhage
  5. Amyloidosis, Wilson's disease
  6. HIV infection

Types

1. **Acute adrenal insufficiency**
   - Shock
   - Fever
   - Abdominal pain, vomiting confused for acute abdomen
     Waterhouse-Friderichson syndrome. It is a bilateral adrenal infarction associated with meningococcal sepsis.
2. **Chronic adrenal insufficiency**
   - Anorexia, weakness, nausea
   - ACTH and POMC—pro-opiomelanocortin levels increase—resulting in hyperpigmentation of skin and oral mucosa
   - Hypotension, hypercalcaemia and hyponatraemia

Diagnosis

- Basal ACTH levels are increased
- Cortisol levels are decreased
- ACTH tests: No rise in cortisol levels following administration of ACTH.

Treatment

- **Acute cases:** IV hydrocortisone 100 mg 6th hourly, saline transfusion, control of infections and cardiac care.
- **Chronic cases:** Oral hydrocortisone (10 mg/m^2 body surface area) and fludrocortisone (0.1 mg)
- Lifelong treatment with glucocorticoid and mineralocorticoid replacement.

I. Neoplasm of the sympathetic neurons

1. **Ganglioneuroma:** It is a benign neuronal tumour, commonly arising from retroperitoneal lumbar sympathetic trunk. FNAC, ultrasound followed by surgical excision is the management.
2. **Neuroblastoma**

II. Neoplasm of the chromaffin cells

- Phaeochromocytoma

NEUROBLASTOMA

- It is a malignant tumour, derived from sympathetic nervous system and adrenal medulla.
- Since it originates from the neural crest, it may be found from **orbit to pelvis** where sympathetic nervous tissue is found.
- It occurs in 1 in 10,000 live births.
- It is the **most common solid tumour** of infancy and childhood.
- Adrenal gland is **the most common** site of neuroblastoma.
- As the name suggests, the tumour occurs due to **malignant proliferation of the neuroblasts** or failure of maturation of primitive sympathetic nerve cells, the neuroblasts (Key Box 20.13).

Pathology

- Characterised by presence of immature cells derived from neuroectoderm of sympathetic nervous system.
- It is pale with grey surface and well-encapsulated.

KEY BOX 20.13

NEUROBLASTOMA

- Most common solid tumour in infancy and childhood.
- Adrenal gland is the most common site.
- Mass abdomen and metastasis are common presenting features.
- Surgical excision is the best treatment.
- Younger the child, better the prognosis.
- **Highest incidence of spontaneous remission.**

KEY BOX 20.14

TUMOURS WITH CALCIFICATION

- Neuroblastoma
- Chondrosarcoma
- Papillary carcinoma thyroid
- Phaeochromocytoma (10%)

- Few areas of calcification are seen (Key Box 20.14).
- Necrosis and haemorrhage are late features.
- Microscopically—uniform round cells containing hyperchromatic speckled nucleus, **Homer-Wright rosettes** with central fibrillar core.

Clinical features

- 50% of children present to the hospital **under the age of one year** and 80–90% are less than 3 years of age.
- An **abdominal mass** is the most common presenting feature. The mass has all the features of a renal mass but location is slightly higher. It is firm to hard, nodular and fixed.
- The child is **sick** with weight loss, fever, abdominal distension, anaemia, etc.
- Functional tumours produce diarrhoea and hypokalaemia due to release of **vasoactive intestinal polypeptide** (VIP), sweating and flushing due to release of **catecholamines**.
- **Proptosis** and **periorbital swellings** are due to bony metastasis and subcutaneous nodules are quite common (Key Box 20.15).
- Secondaries in retro-orbital region results in infraorbital ecchymosis—**Raccoon's eye sign.**
- Posterior mediastinal neuroblastomas can produce cord compression and even paraplegia due to protrusion within the spinal canal **(dumbbell tumours).**

Investigations

- **Vanillyl mandelic acid (VMA)** and homovanillic acid (HVA) are the byproducts of catecholamines passed in the urine. 24-hour urinary excretion of catecholamines and these two metabolites will be very high.
- **Plain X-ray** abdomen shows fine stippled calcification. X-ray chest is done to rule out cannon-ball secondaries.
- Abdominal **ultrasound** and **computed tomography (CT)** can define the mass, nature and extent of the tumour and can detect hepatic metastasis.

KEY BOX 20.15

TYPES OF NEUROBLASTOMA

1. Pepper type: Right-sided tumours with secondaries in the liver.
2. Hutchinson's type: Left-sided tumour with metastasis in bones—orbit and skull.

- **Magnetic resonance imaging (MRI)** is better than CT scan in detecting the mass as well as bony metastasis. CT/MRI guided core biopsy can be done.
- **Bone marrow aspiration** is positive in around 60–70% of cases.

Treatment

- **Intermediate and high risk** cases can be managed by chemotherapy followed by surgery.
- **Early cases** respond very well to surgical excision.
- However, many children present with metastasis and, chemotherapy and radiotherapy are given first to control the disease followed by surgical excision. **Autologous bone marrow transplantation** has improved the outlook for patients with advanced disease.

PHAEOCHROMOCYTOMA

Definition: These are catecholamine producing tumours arising from adrenal medulla and sympathetic ganglia which are derived from chromaffin cells (Key Box 20.16).

- As the name suggests, phaeo (dusty) chromo (chromaffin cells)—phaeochromocytoma is a neoplasm arising from chromaffin cells.
- 90% of the tumour occurs in adrenal glands.
- When phaeochromocytoma occurs in extra-adrenal sites, it is called paraganglioma.
- Extra-adrenal sites include organ of Zuckerkandl (the most common site), urinary bladder, renal hilum, chest, neck, etc. These are the sites of paraganglionic system.
- In about 5% of cases, the tumour can be a component of multiple endocrine neoplasia (MEN) Type IIa or Type IIb.
- Other syndrome associated with **Phaeochromocytoma is von Hippel-Lindau syndrome** (Key Box 20.17).
- The clinical manifestations are due to release of adrenaline and noradrenaline. When the level of noradrenaline is high, symptoms are severe.

Pathology

- It is a soft, highly vascular tumour consisting of large sympathetic ganglionic cells. Most of the cells are differentiated.
- Usually, it is small in size and well-encapsulated.
- Sometimes, it can present as a large abdominal mass.

KEY BOX 20.16

PHAEOCHROMOCYTOMA

10%	Bilateral
10%	Malignant
10%	Extra-adrenal
10%	Multiple
10%	Familial
10%	Children

This 10% rule has been challenged

KEY BOX 20.17

OTHER NEUROECTODERMAL ANOMALIES WITH PHAEOCHROMOCYTOMA

- Neurofibromatosis (NF) type I
- Acoustic neuroma
- Meningioma, glioma, haemangioblastoma (von Hippel-Lindau (VHL) syndrome
- Astrocytoma
- MEN type II—hereditary phaeochromocytoma
- Familial paraganglioma syndrome along with carotid body tumours

- Microscopic features are **polygonal** or spheroidal chromaffin cells within a vascularised fibrous stroma.
- Presence of high number of Ki-67 positive cells, vascular invasion or breached capsule and PASS—phaeochromocytoma of the adrenal gland scale score suggests metastasis.

Clinical features

- Sporadic cases peak at 40–50 years, whereas familial tend to occur early.
- The most common presenting feature is paroxysmal or persistent hypertension (80–90%). It is associated with palpitation (50–70%), fever, pallor, tremors, sweating and severe headache (60–90%).
- The paroxysmal attack may last for a few minutes to a few hours (Key Box 20.18).
- **The factors which stimulate an attack are**:
  - Surgery
  - Anaesthesia
  - Invasive procedure
  - Late pregnancy
  - Drugs—Histamine, glucagon, etc.
  - Palpation of the mass
  - **Classical triad** consisting of headache, diaphoresis and palpitations is seen in only a few patients.
- **A high index of suspicion is necessary to diagnose phaeochromocytoma in a hypertensive patient.**

Investigations

1. **Urinary levels of free catecholamines**, vanillyl mandelic acid (VMA) in excess of 7 mg/24 hours and meta-adrenaline 1–3 mg/24 hours are suggestive of phaeochromocytoma.

KEY BOX 20.18

HYPERTENSION IN PHAEOCHROMOCYTOMA

- **P**aroxysmal
- **P**alpitations
- **P**rofuse sweating
- **P**alpation of the mass
- **P**ersistent
- **P**allor
- **P**ain abdomen

Observe **7 Ps**

2. **Plasma metanephrines:** A fractionated plasma free metanephrine level may be measured. Blood sample drawn after 15–20 minutes of IV catheter insertion (not immediately)
3. **Computed tomography**:
   - Noninvasive, safe investigation
   - It has a high degree of accuracy
   - It can pick up lesions of less than 1 cm in size.
4. **MRI** is preferred because **contrast media used for CT scan can provoke paroxysms.**
5. ^{131}I-Meta-Iodo-Benzyl-Guanidine (MIBG) scan
   - Iodine labelled MIBG (radionuclide) scan is found to be very specific for phaeochromocytoma. This radionuclide scan locates only abnormal adrenal tissue and is **more useful in detecting ectopic sites of phaeochromocytoma** (Figs 20.14).

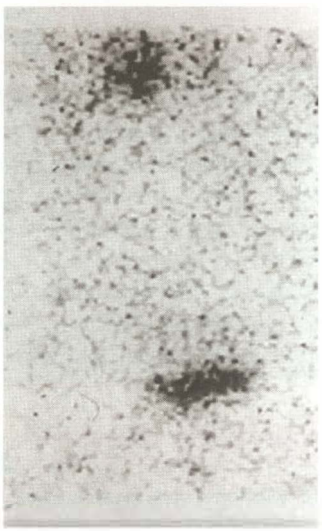


Fig. 20.14: MIBG scan showing extra-adrenal phaeochromocytoma

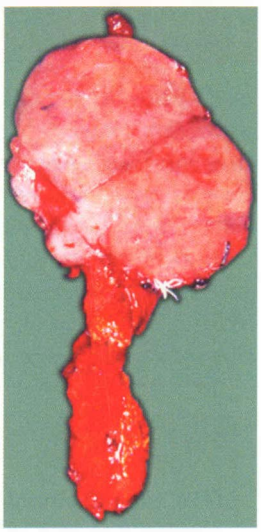


Fig. 20.15: Phaeochromocytoma excised

(*Courtesy:* Prof MG Shenoy, Senior Consultant in Surgery, KMC, Manipal)

Treatment (Fig. 20.15 and 20.16)

- Surgical removal is the treatment[1]. However, a good preoperative preparation is essential before doing surgery. It includes control of blood pressure and tachycardia, by α and β-blockers (Key Box 20.19).
- Contraction **of vascular bed which occurs** due to **catecholamines results in hypovolaemia** which must **be corrected with IV fluids**.
- **First**, alpha-adrenergic blockers such as phenoxybenzamine is given to control hypertension and to permit **re-expansion of intravascular volume**. **Only after the complete alpha-adrenergic blockade, beta-adrenergic blockade may be added**.

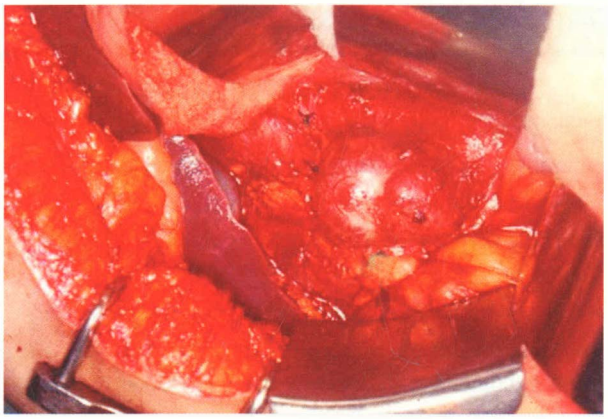


Fig. 20.16: Phaeochromocytoma at surgery

KEY BOX 20.19

PHAEOCHROMOCYTOMA (PREOPERATIVE PREPARATION)

- **Phenoxybenzamine:** 20–60 mg/day for 3–4 weeks before surgery to control **hypertension**—increased to 10 mg/day—total of 100–160 mg.
- **Propranolol:** 20–60 mg/day for 5–7 days before surgery to control **tachycardia** and **arrhythmias**.
- Plenty of **fluids** before surgery to correct **hypovolaemia**.

TEN COMMANDMENTS OF PHAEOCHROMOCYTOMA

1. MRI is better because CT scan provokes paroxysm
2. MIBG—^{123}I single photon emission CT will diagnose about 90% of cases
3. Should not do FNAC/biopsy of adrenals
4. **α-adrenergic blockers** should be started first
5. β-blockade is always given **after alpha blockade**
6. CVP and intra-arterial monitoring is a must, as they may hold a large volume of fluids
7. Should also look on the contralateral side for phaeochromocytoma
8. Ligate adrenal vein first
9. Never allow vaginal delivery in patients who have phaeochromocytoma
10. Laparoscopic adrenalectomy is the 'Gold standard'

Surgery (*see* ten commandments given above)

Excision is the treatment. Certain steps of excision (adrenalectomy) are as follows (Key Box 20.20):

- **Sodium nitroprusside** must be kept ready to treat hypertension during surgery as it is a **direct arterial vasodilator** (0.5 to 10 μg/kg/minute).
- Thorough search in the abdomen **for other sites**, due to the possibility of multiple tumours.

[1]Surgery for phaeochromocytoma is like going to a war. Without proper preparation, going to war is a sure failure.

KEY BOX 2.20

PRECAUTIONS DURING SURGERY

- Midline incision for familial cases
- Anterior or posterior approach for sporadic cases
- Careful positioning
- **Ligation of adrenal vein first**
- Gentle handling
- Haemodynamic monitoring—CVP, arterial monitoring
- Avoid rupture of the tumour to prevent recurrence
- Laparoscopic adrenalectomy is very popular now

- **Postoperative hypotension** can be a serious problem which needs to be treated with large volume of plasma expanders, blood transfusions, corticosteroids and vasopressors.

MALIGNANT PHAEOCHROMOCYTOMA

- 10% of phaeochromocytomas are malignant.
- Rate is higher in extra-adrenal tumours
- Malignancy is suspected when metastasis or vascular invasion is present or when a capsule is breached
- Open adrenalectomy is the choice for excision.
- Debulking should be done even if metastasis is present, so as to decrease tumour burden and to control catecholamine excess.

MISCELLANEOUS

INCIDENTALOMAS

- Incidentally detected adrenal masses.
- Detected by ultrasound or CT scan (1 to 5% of patients undergoing CT abdomen for evaluation of abdominal pain).
- Some patients may have subclinical Cushing's syndrome or aldosteronism.
- Nonfunctioning situations may be metastatic cancer (primary from breast, kidney, melanoma, lung)
- Myelolipoma
- Adrenal cysts
- Majority of such tumours are **nonfunctioning cortical adenomas of no clinical significance**.
- Indication to do adrenal gland biopsy is to rule out/diagnose metastasis
- Absolute contraindication to do adrenal biopsy in phaeochromocytoma
- Any nonfunctioning adrenal incidentaloma should be resected
- Majority of incidentalomas are nonfunctional adenomas.

See clinical notes.

CLINICAL NOTES

An 18-year-old girl was admitted for tonsillectomy. This girl suffered from occasional headache. She was diagnosed to have migraine. Preoperative blood pressure showed mild elevations which were thought to be due to anxiety. During tonsillectomy, there was a rise in BP, which was controlled well. However, in the postoperative period, there was severe tachycardia, hypertension, arrhythmias and hypovolaemia. Within 8 hours of surgery, the patient died even as the diagnosis of phaeochromocytoma was being considered.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

New key boxes and pearls of wisdom have been added. Disorders of adrenal cortex including Conn's syndrome and Cushing's syndrome have been added.

MULTIPLE CHOICE QUESTIONS

1. Superior parathyroids derive blood supply from:
A. Common carotid artery
B. Internal carotid artery
C. Inferior thyroid artery
D. Superior thyroid artery

2. Superior parathyroids are found:
A. Above inferior thyroid artery
B. Below inferior thyroid artery
C. Above superior thyroid artery
D. Below superior thyroid artery

3. Following are true for primary hyperparathyroidism *except:*
A. Majority are due to a single adenoma
B. Majority are sporadic
C. Hypercalcaemia with decreased PTH levels are often seen
D. Rarely it can be due to carcinoma also

4. Following are true for inferior parathyroid glands *except:*
A. Derived from 3rd pharyngeal pouch
B. It is inferior to inferior parathyroid artery
C. It is superior to inferior parathyroid artery
D. Inferior parathyroids are less constant in position

5. Following are functions of parathyroid gland *except:*
A. Activates osteoclasts to resorb bone
B. Increased calcium reabsorption from urine
C. Renal activation of vitamin D
D. Decrease renal excretion of phosphate

6. Following is not the feature of primary hyperparathyroidism:
A. Hypercalcaemia
B. Most common manifestation of MEN type I
C. Band keratopathy in cornea
D. Dystrophic calcification

7. The best investigation of choice to localise parathyroid gland is:
A. ^{99m}Tc-labelled sestamibi scan
B. CT scan
C. MRI
D. Ultrasound

8. Which one of the following is not the site of inferior parathyroid gland:
A. Thyrothymic axis
B. Mediastinum
C. Within carotid sheath
D. Cricothyroid articulation

9. Following are true for parathyroid hyperplasia *except:*
A. It affects all 4 glands
B. It is not the common cause of hyperparathyroidism
C. Total parathyroidectomy is the treatment of choice
D. Thymectomy need not be done

10. Which one of the following is the treatment of choice for acute hypercalcaemic crisis?
A. Steroids
B. Diuretics
C. Vasopressors
D. Bisphosphonates

11. Which is the first drug to used in the preoperative preparation of a patient with phaeochromocytoma?
A. Steroids
B. Noradrenaline
C. Propranalol
D. Phenylbenzamine

12. Which one of these operative steps is done first in excision of phaeochromocytoma?
A. Ligation of adrenal vein
B. Ligation of inferior phraenic artery
C. Ligation of branch of renal artery
D. Isolation from kidney

13. A few landmarks for identification of ectopic sites of phaeochromocytoma include following *except*:
A. Urinary bladder
B. Along 5th cranial nerve
C. Organ of Zuckerkandl
D. Superior para-aortic region—below the diaphragm and renal poles

14. Following are true for management of phaeochromocytoma *except*:
A. Phenoxybenzamine is given prior to propranolol
B. Intra-arterial monitoring is a must
C. It is dangerous to infuse large volumes of fluid after ligation of adrenal vein
D. Laparoscopic resection is the gold standard

15. Following are true for right adrenal gland *except*:
A. Right adrenal gland drains into renal vein
B. Right adrenal gland is partly behind inferior vena cava
C. Right adrenal gland is pyramidal in shape
D. Right adrenal gland is anterolateral to the right crus

16. The majority of the sporadic primary hyperparathyroidism is due to:
A. Hyperplasia
B. Adenoma
C. Carcinoma
D. Cystic degeneration

17. Severe bone disease due to primary hyperparathyroidism is called as:
A. Sipple syndrome
B. Werner syndrome
C. von Recklinghausen's disease
D. Paget's disease

18. Superior parathyroid gland can be found by:
A. Ligating the middle thyroid vein and dividing the lobe
B. Ligating inferior thyroid artery and mobilising the gland
C. Ligating superior thyroid pedicle and mobilising the gland anteriorly
D. Ligating inferior thyroid veins and diving isthmus

19. Following are locations of inferior parathyroid glan *except:*
A. Upper horn of thymus
B. Within carotid sheath
C. Within thyroid lobe
D. Behind superior thyroid pedicle

20. Following are causes of secondary hyperpara thyroidism *except:*
A. Malabsorption
B. Vitamin D-deficient rickets
C. Pseudohypoparathyroidism
D. Acute renal failure

ANSWERS

1 C	2 A	3 C	4 B	5 D	6 D	7 A	8 D	9 D	10 D
11 D	12 A	13 B	14 C	15 A	16 B	17 C	18 C	19 D	20 D

21

Breast

- Congenital anomalies
- Surgical anatomy
- Cystic swellings of breast—classification
- Acute bacterial mastitis
- Antibioma
- Retromammary abscess
- Phylloides tumour
- Intracystic carcinoma of breast
- Aberrations of normal development and involution
- Fibroadenoma
- Duct ectasia—plasma cell mastitis
- Idiopathic granulomatous mastitis
- Macrocysts
- Galactocoele
- Galactorrhoea
- Duct papilloma
- Carcinoma of the breast
- Effects of lymphatic obstruction from carcinoma breast
- Breast reconstruction
- Carcinoma of male breast
- Prophylactic mastectomy
- Mondor's disease
- Angiosarcoma breast
- Disorders of augmentation mammoplasty
- Rare breast cancers
- A case of carcinoma breast
- Recent changes in the treatment of carcinoma breast
- What is new?/Recent advances

Embryology

- At 5th or 6th week of intrauterine life, 2 ventral bands of thickened ectoderm called mammary ridge/milk line or line of Schultz appear. The line extends from axilla to the groin. Thus, accessory nipples can appear along the milk line from axilla to groin (Fig. 21.1).
- Persistent part of mammary ridge is converted into a pit. Secondary buds develop, they divide and form more lobes.
- The nipple is everted at the site of original position.
- Oestrogens cause enlargement of mammary glands at puberty and progesterone stimulates development of secondary alveoli.

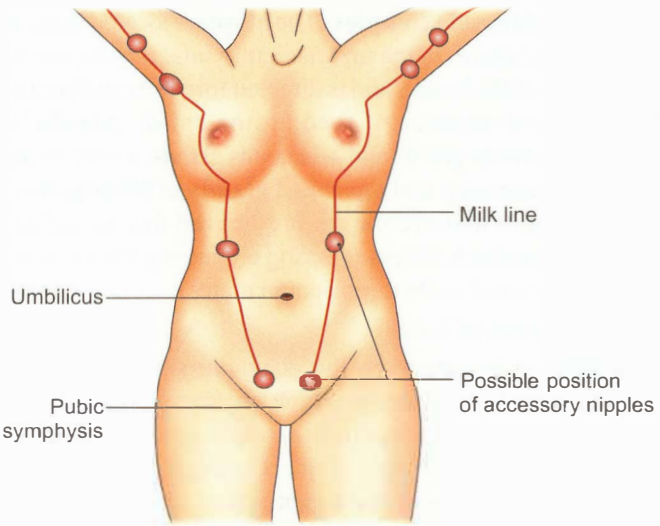


Fig. 21.1: Milk line with possible positions of accessory nipples

CONGENITAL ANOLAMIES OF BREAST

I. **Amazia:** Congenital absence of breast is very rare. It can be unilateral or bilateral.

II. **Poland's syndrome**
- Amazia
- Absence of sternal portion of pectoralis major
- Occurs commonly in males

III. **Supernumerary nipples:** Accessory nipples, polythelia (Fig. 21.2)

IV. **Athelia:** Absence of nipple

V. **Polymastia:** Accessory breasts occur along the milk line—axilla (most common), groin, thigh or buttock (Figs 21.3 and 21.4).

VI. **Micromastia:** Due to congenital defects of ovary, lack of hormonal stimulation occurs which results in small breast.

Fig. 21.2: Accessory nipple

Figs 21.3 and 21.4: Accessory breasts

SURGICAL ANATOMY OF BREAST

- Breast, a modified sweat gland occupies the pectoral region from the 2nd to the 6th rib vertically, and from the lateral border of sternum to the midaxillary line, horizontally. It is hemispherical, and lies in the superficial fascial planes.
- It is composed of fatty tissue and does the function of secreting milk. **The axillary tail of Spence is the part of the breast which is in the axilla and is deeper to the deep fascia**, whereas the entire breast is a *subcutaneous structure*.

Structure of the breast (Fig. 21.5)

1. **Nipple and areola complex:** The **nipple** is located in the 4th intercostal space, in the midclavicular line. It is the erectile structure of the breast, and is directed forwards and laterally for the convenience of feeding the child. **Areola** has modified sweat glands and sebaceous glands. These enlarge during pregnancy and are called **glands of Montgomery**. Both nipple and areola are pigmented due to melanin deposition which increases during pregnancy. Hair is absent in the areola of women (present in males).
2. **Parenchyma** of breast.
3. **Stroma** gives support to the glandular structure. Therein lie **ligaments of Cooper** which are cone-shaped fibrous bands. Their apex is attached to overlying skin, and base to the fascia over pectoralis major. **Puckering of the skin is due to infiltration of the ligaments of Cooper.**
4. **Lobule** is the chief functional and structural unit of breast. Many lobules join to form a lobe. There are **15–20 lobes and** each lobe is drained by a **lactiferous duct**. They are 15–20 in number arranged radially, lined by myoepithelial cells, which converge into the nipple. Diameter of a lactiferous duct is 2–4 mm.

Lymphatic drainage

They can be divided into lymph nodes and lymphatics (Fig. 21.6).

I. Lymph nodes

1. Anterior or pectoral: They are under the pectoralis major which forms anterior fold of axilla

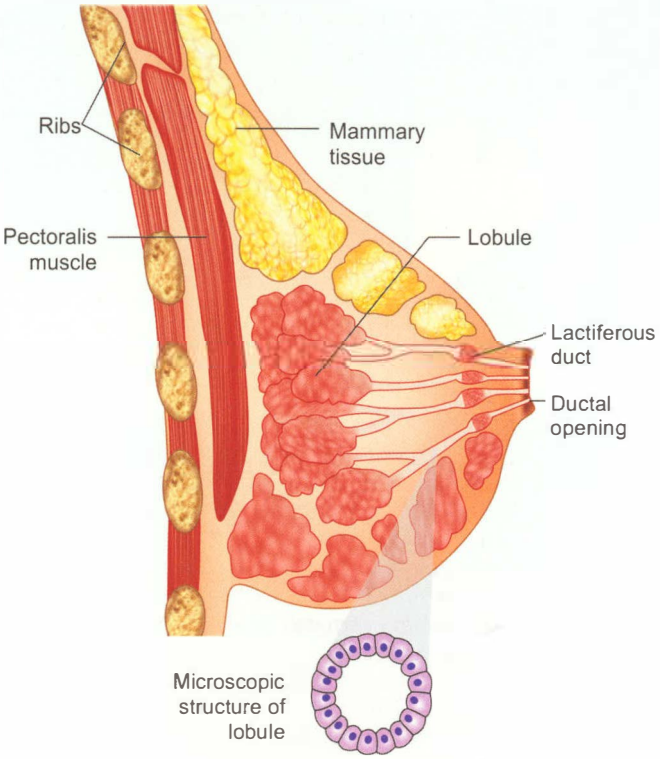


Fig. 21.5: Structure of the breast

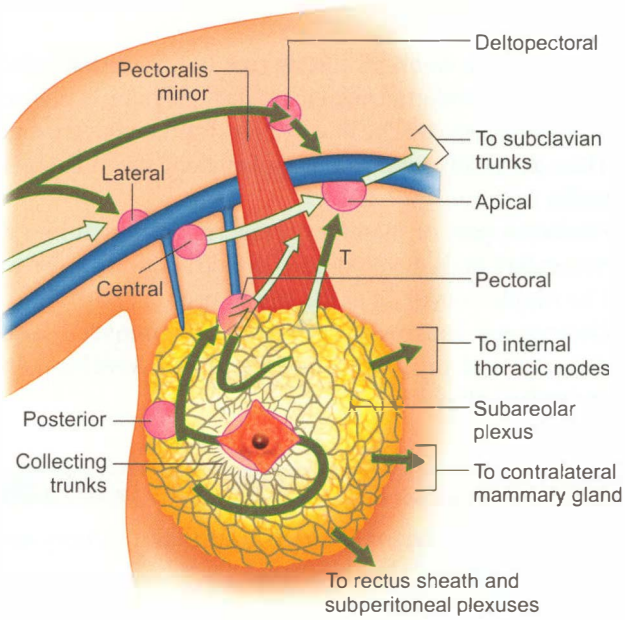


Fig. 21.6: Lymph nodes and lymphatic spread from breast

2. Central group: These lymph nodes are present in the centre of axilla (arm pit). One has to dip the examining fingers slightly deeper into axilla to detect the enlargement.
   - Anterior and central group of nodes are commonly involved in carcinoma breast.
3. Lateral group are felt against humerus. They are also called brachial group.
4. Apical: They are also called infraclavicular nodes, situated very high in the axilla. They are difficult to feel clinically.

5. Posterior: They are also called subscapular group of lymph nodes. They are felt along the posterior fold of the axilla. These five groups together form the axillary group of lymph nodes.
6. Internal mammary lymph nodes: Also called parasternal nodes. They lie along internal mammary vessels. They are located in the 2nd, 3rd and 4th space.
7. Supraclavicular lymph nodes: Spread to supraclavicular lymph nodes indicate advanced stage of the disease. It indicates poor prognosis.

Miscellaneous lymph nodes/plexus

1. Cephalic nodes—Deltopectoral nodes
2. Interpectoral nodes—Rotter's nodes
3. Posterior intercostal nodes—in front of heads of ribs
4. Intra-abdominal—Subdiaphragmatic/retroperitoneal.

II. Lymphatic Vessels

1. **Superficial lymphatics:** Drain skin over the breast except nipple and areola. Superficial lymphatics of one breast communicate with the contralateral breast across midline.
2. **Deep lymphatics:** Drain parenchyma of the breast. They also drain nipple and areola.

Important key points about lymphatics and spread

- The first **lymph nodes** draining the tumour bearing area is called **sentinel node**.
- 75% of the lymph from the breast drains into axillary nodes.
- 20% drains into the internal mammary nodes
- 5% of lymph drains into posterior intercostal lymph nodes.
- Most of the lymphatics eventually drains into central to apical and then to supraclavicular lymph nodes.
- Internal mammary nodes receive lymphatics not only from inner quadrant but also from outer quadrant.
- Lymphatics from inner quadrant of the breast penetrate rectus sheath and thus spread into coelomic cavity. It results in ascites, rectovesical deposits and Krukenberg tumours.
- Krukenberg tumours are bilateral bulky ovarian metastasis in premenopausal women. During ovulation, raw surface develops over the ovary into which malignant cells drop and develop into large tumours (transcoelomic spread).

Blood supply of the breast (branches of axillary artery)

1. **Lateral thoracic artery** gives many branches which penetrate through the pectoralis major and supply the breast.
2. **Internal mammary artery** gives branches which perforate intercostal spaces.
3. **Pectoral branches of thoraco-acromial artery** supply upper part of the breast.
4. **Lateral branches of posterior intercostal arteries**

Veins: Breast is drained by perforating branches of internal mammary veins, tributaries of axillary veins and perforating branches of posterior intercostal veins.

Venous return follows the arteries but drain into large veins that also receive blood from vertebrae and thoracic cage. E.g. posterior intercostal veins joining paravertebral plexus of veins (Batson's venous plexus). This explains the occurrence of metastasis in the vertebrae and pelvic bones from carcinoma of the breast.

PHYSIOLOGY (Key Box 21.1)

KEY BOX 21.1

PHYSIOLOGY OF THE BREAST

- ***Oestrogen:*** Initiates ductal development.
- ***Progesterone:*** Differentiation of epithelium and lobular development (glandular development)
- ***Prolactin:*** Lactogenesis in late pregnancy and postpartum period. It also upregulates hormone receptors and stimulates epithelial development.

CYSTIC SWELLINGS OF BREAST

Classification

1. **Inflammatory:** Acute bacterial mastitis with abscess*
2. **Neoplastic:**
   a. ***Benign:*** Phylloides tumour*
   b. ***Malignant:*** Intracystic carcinoma*
3. **Non-neoplastic cyst**
   a. Fibroadenosis—cyclical mastalgia
   b. Simple cysts of the breast
   c. Cyst of Bloodgood—blue domed cyst
4. **Retention cyst of the breast:** Galactocoele
5. **Other rare causes of cysts of the breast**
   a. Tuberculous mastitis with cold abscess*
   b. Lymphatic cyst of the breast (congenital)
   c. Hydatid cyst of the breast
   d. Haematoma of the breast

ACUTE BACTERIAL MASTITIS (BREAST ABSCESS— PYOGENIC MASTITIS)

Aetiopathogenesis

1. Lactational mastitis

It is most commonly encountered during **lactation.** Hence, it is called lactational mastitis.

Precipitating factors

- Crack/fissure in the nipple
- Retracted nipple. Hence, cleaning of the breast is a problem
- Oral cavity infection in the child

*These swellings may give rise to fluctuation in a 'small' part of the swelling. Truly, they are not cystic swellings.

Fig. 21.7: Large breast abscess presented late to the hospital. Managed by incision and drainage (I and D), not aspiration

Fig. 21.8: Extensive necrosis of skin due to severe mastitis (*Courtesy:* Dr C.G. Narasimhan, Surgeon, Mysore, Karnataka)

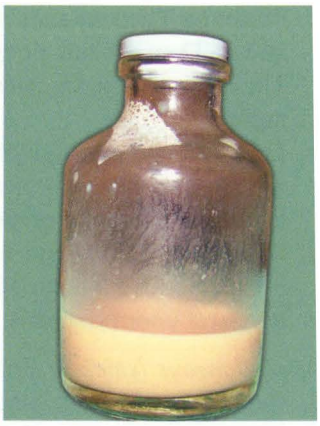


Fig. 21.9: Current thinking is that repeated aspiration of the breast abscess can also be an alternative treatment to incision and drainage

2. Haematoma

- Infection in a haematoma can result in an abscess—rare cause.
- Staphylococcus produces many enzymes/toxins such as catalase, coagulase, hyaluronidase which result in an abscess. It also inhibits phagocytosis because of type 'A' protein on its surface. *Staphylococcus aureus,* which enters through the nipple, proliferates intraductally and produces *clotting of the milk.* Within the clot the organisms multiply, which results in a **cellulitic stage of the breast** (mastitis) and in untreated cases, it may give rise to a breast abscess. Initially, only one lobule and duct get affected. Later other lobules get infected, giving rise to an intramammary abscess.

3. Nonlactational breast abscess

- It occurs in patients with duct ectasia and periductal mastitis. When such an abscess ruptures, it results in a mammary duct fistula. It classically drains at the junction between the areola and breast skin. **Anaerobic bacteria** are the cause in majority of cases.

4. **Other factors:** Diabetes, AIDS and chronic illness also can give rise to breast abscess (Key Boxes 21.2 and 21.3).

Clinical features

- Severe pain in the breast due to spreading inflammatory exudate. Breast is swollen, tense, tender and warm to touch. These are the signs of *cellulitic stage.*
- Once *breast abscess* develops, there is high grade fever with chills and rigors and a soft, cystic fluctuant swelling can be felt in the breast. In untreated cases, abscess may rupture through the skin resulting in *necrosis of the skin of the breast, ulceration* and discharge (Figs 21.7 and 21.8)
- In deep-seated abscess, it is difficult to elicit fluctuation and often fluctuation is a late sign. Hence, if **throbbing pain and fever with chills and rigors** are present, immediate drainage is mandatory. If not done, significant amount of breast tissue will be destroyed.

Treatment (Key Box 21.4)

- **Stage of cellulitis**
  - Not to feed the child on the affected side.
  - Cloxacillin 500 mg, 6th hourly, orally for 7–10 days.
  - Anti-inflammatory drugs such as ibuprofen 400 mg, three times a day.
  - Good support to the breast.

KEY BOX 21.2

MRSA AND BREAST ABSCESS

- Methicillin resistant *Staphylococcus aureus* (MRSA) or community acquired MRSA or CA-MRSA can cause breast abscess in patients who have no traditional risk factors
- Hospitals, nursing homes (patients with open wounds) are the risk areas
- Co-amoxyclav 1000 mg 2 times/day—7–10 days
- Erythromycin 500 mg 3 times/day—7–10 days
- Vancomycin 1.5 g vial 12th hourly x 7–10 days
- Suspect MRSA infection when abscesses recur, abscess persists and is nonlactational.

KEY BOX 21.3

BREAST ABSCESS

- Common organism—*Staphylococcus aureus*
- Retracted nipple is one of the causes
- Commonly seen during **lactational period**
- Very painful condition
- Do not wait for fluctuation
- Guided aspiration should be done (Fig. 21.9)
- Ultrasound can be done
- Cloxacillin is the drug of choice
- Nonlactational abscess—Metronidazole is drug of choice

Fig. 21.10: Nipple retraction—she also had a lump-antibioma. Observe previous drainage incision

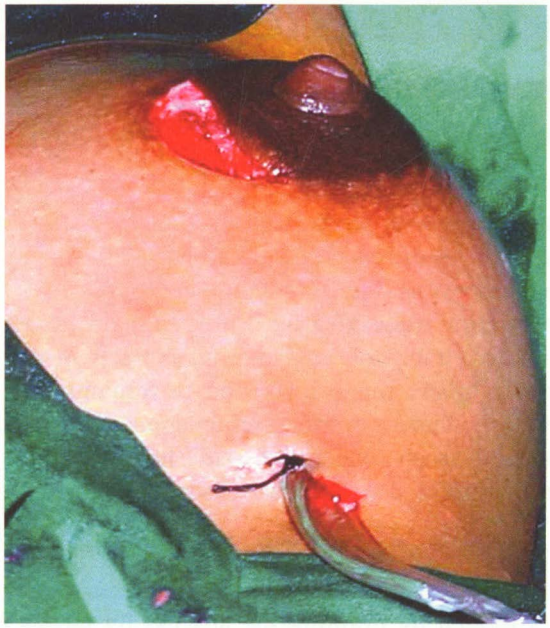


Fig. 21.11: Breast abscess drainage. Observe corrugated drainage tube. Mostly it is replaced by tube drains today

KEY BOX 21.4

TREATMENT OF BREAST ABSCESS

- Pus is frequently expressed from nipple
- Breast feeding is stopped
- Antibiotics are started
- **Repeated aspiration** with antibiotic cover with or without ultrasound guidance can be offered (Fig. 21.9)
- Often abscess is confined to one sector. Hence, aspiration can be advised.
- Incision and drainage is also an accepted treatment, more so in large abscesses with purulent discharge (Fig. 21.11).
- Large, brawny, sterile oedematous swelling that occurs after antibiotic therapy is ANTIBIOMA (Fig. 21.10).

- For nonlactational breast abscess, add metronidazole 400 mg, 3 times a day for 5–7 days.

- **Stage of abscess**
  - The abscess should be drained—incision and drainage (I and D) under antibiotic cover.
  - If the abscess is situated in any quadrant of breast, other than lower quadrant, it is drained by **radial incision.**
  - Abscess in lower quadrant is drained by **inframammary** incision placed in the inferior aspect of breast (*refer to* breast abscess drainage under operative surgery section).
  - When both the breasts have an abscess, the breasts should be emptied and the milk that is expressed *can be boiled and given to the child.*

PEARLS OF WISDOM

Currently, the initial treatment policy is USG-guided drainage.

Complications of acute mastitis (Key Box 21.5)

KEY BOX 21.5

COMPLICATIONS OF ACUTE MASTITIS

- Abscess
- Skin necrosis
- Toxaemia
- Antibioma

CLINICAL NOTES

An 18-year-old girl was admitted in a medical ward with the diagnosis of pyrexia of unknown origin (PUO). All investigations were normal except a high total WBC count. After about a week, the duty nurse noticed that the patient's dress was stained with pus. It was a large breast abscess with necrosis of the overlying skin. It was drained later. The girl was so shy that she did not even complain of pain or swelling of the breast. These types of cases are not uncommon in our country.

ANTIBIOMA

- It means an antibiotic induced swelling. (Oma) = Tumour (swelling).
- When an abscess occurs in the breast and antibiotics are given, without draining the abscess, the abscess cavity may become fibrous and it results in **firm to hard lump** in the breast. It gives rise to vague **ill health of the patient**.
- This hard lump can be confused for *malignancy.*
- It is treated by excision.

OTHER TYPES OF BREAST ABSCESSES

RETROMAMMARY ABSCESS

It is collection of pus behind the *pectoralis major* (Fig. 21.12).

Common causes

1. Haematoma with secondary infection.
2. Tuberculosis of ribs with cold abscess.
3. Cold abscess arising from lymph node.
4. **Empyema necessitans:** Empyema of lung, if left untreated, tracks out and the pus collects in the subcutaneous plane posteriorly and retromammary region anteriorly, thereby forming retromammary abscess. There may be a tense, tender and cystic lump palpable which can be confused with breast abscess.

Management

- Chest X-ray to rule out pulmonary tuberculosis.
- It is treated by draining the abscess by means of submammary (Galliard-Thomas) incision.

SUBAREOLAR ABSCESS

- It is common in nonlactating women.
- It communicates with lactiferous duct resulting in mammary fistula.
- In chronic cases, retraction of the nipple can occur which is partial or slit-like.
- It can also be due to an infected sebaceous cyst.

TUBERCULOUS MASTITIS

- Incidence is 1 to 4% in India
- Poor socioeconomic conditions

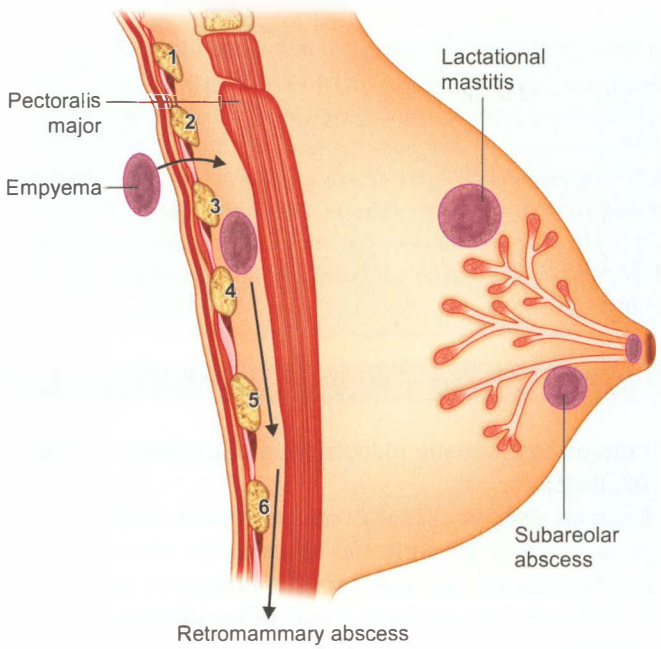


Fig. 21.12: Different types of breast abscess

- Presents as a lump, which can be hard and mimic carcinoma
- More common in the reproductive age group—mainly in lactating women.
- Clinical features may include multiple tender abscesses sinuses and matted nodes in the axilla (need not always be present).
- Involvement of nipple and areola is very rare.
- It is almost secondary to a pulmonary lesion—retrograde lymphatic spread via paratracheal and internal mammary lymph nodes.
- **Few types:** Nodular, sclerosing, disseminated type, TB mastitis obliterans and miliary form.
- FNAC, biopsy will help in the diagnosis—caseating granuloma with Langhan type of giant cells.
- Aspiration of pus, lumpectomy, excision/even mastectomy may be required along with concurrent antitubercular treatment (Fig. 21.13).

Fig. 21.13: Tuberculous mastitis *(Courtesy:* Dr Maruthu Pandyan, Govt Medical College, Madurai)

PHYLLOIDES TUMOUR

- They are fibroepithelial tumours composed of an epithelial and cellular stromal component.
- Earlier known as cystosarcoma phylloides and mistakenly labelled as a giant intracanalicular fibroadenoma. They are tumours of the breast, with a predilection to attain massive size and recur locally after lumpectomy (Figs 21.14 to 21.18).
- Account for less than 1% of all breast neoplasms

Types of phylloides

- Benign: **More than 60% younger women**
- Borderline Depends upon mitotic activity, cellularity
- Malignant and infiltration at the edge of the tumour

Clinical features (Key Box 21.6)

- Age group—30 to 40 years—premenopausal
- Rapid growth
- Stretched, shiny skin
- Red, dilated veins over surface, warm to touch
- Bosselated surface (big nodules), a few cystic areas.

It is differentiated from carcinoma by:

1. No fixity to the skin
2. No fixity to the pectoralis—mobile on the chest wall
3. Lymph nodes will not be involved
4. No nipple retraction.

KEY BOX 21.6

BOSSELATED SURFACE—CONDITIONS

- Phylloides tumour
- Polycystic kidney
- Polycystic liver
- Large nodular goitre

- As the tumour grows very fast, it undergoes necrosis in various places resulting in cystic areas within the breast. It discharges serous fluid. Hence, the name *serocystic disease of Brodie. However, it rarely feels cystic.*
- Histologically, the tumour cells have a branching pattern, penetrating the cystic cavity *(phyllus* means leaf-like pattern). Fibrous stromal proliferation is a feature.

Diagnosis

- Very often, it is a clinical diagnosis
- Ultrasound will help to detect the size of the tumour, solid and cystic areas
- Trucut biopsy may reveal mitotic figures suggestive of malignancy

Treatment

- Small: Wide excision (1–2 cm margin). Lumpectomy should not be done as it can cause recurrence.
- Giant. More wider excision
- Malignant: More wide excision. Sometimes, simple mastectomy may be necessary.

Fig. 21.14: Phylloides tumour of the left breast causing massive enlargement

Fig. 21.15: Phylloides tumour of the left breast at surgery

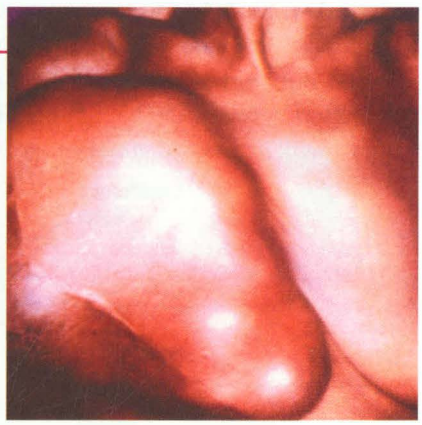


Fig. 21.16: Malignant phylloides tumour (recurrent)

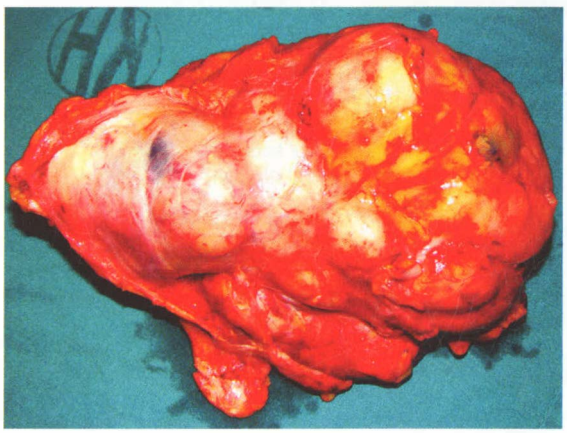


Fig. 21.17: Phylloides tumour of the left breast at surgery (*Courtesy:* Dr Hartimath B, Associate Professor, KMC, Manipal)

Fig. 21.18: Phylloides tumour with dilated veins (*Courtesy:* Dr Sreejayan, Professor of Surgery, Calicut Medical College, Kerala)

Complications

1. Pressure necrosis—secondary infection
2. Distant spread—lungs and long bones

Intracystic carcinoma of breast: Reeclin's disease

Rapidly growing carcinoma of the breast can undergo cystic degeneration resulting in a cyst, called intracystic carcinoma of the breast. Aspiration reveals haemorrhagic fluid, containing malignant cells and which refills after aspiration. Other features of carcinoma of the breast may be present.

Normal process

Aberration
(Benign breast disorder)
Conditions considered too common an occurrence and too mild a state to warrant the designation of a diseased state

Disease
(Benign breast disease)

(*see* Table 21.1 and 21.2)

ABERRATIONS OF NORMAL DEVELOPMENT AND INVOLUTION OF THE BREAST

The concept of Aberrations of Normal Development and Involution (ANDI) of the breast was first published by **LE Hughes** et al of **Cardiff Breast Clinic** in **1987**. The ANDI classification is based on pathogenesis, and recognises that a spectrum exists from normal, through mild abnormality, to disease. This has resulted in a radical change in attitude to the understanding and management of breast disorders. Changes previously regarded as disease are so common that they must be regarded as lying within the spectrum of normality (Fig. 21.19).

This concept is of value in dispelling the supposed association between the benign conditions and cancer. Most patients with lumps and mastalgia are concerned that they may be harbouring cancer (the result of a successful information campaign!!), and once a definite opinion of the benign nature of the lump is conveyed and the patient is informed that no further treatment is required, most would be reassured.

Having noted the above, a word of caution is due here—It is preferable to overtreat a benign breast disease rather than miss or delay treatment of an early carcinoma of the breast (Fig. 21.19). So following the clinical dictum "In a patient who is above the age of 40, with a recently detected lump in the breast, it should be considered to be carcinoma breast until proved otherwise" may prove to be a wrong answer in many but the correct management in most.

From the above, it follows that, in case of doubt (where there is a high risk patient or doubtful signs of malignancy) it would be prudent on the part of the treating surgeon to rule

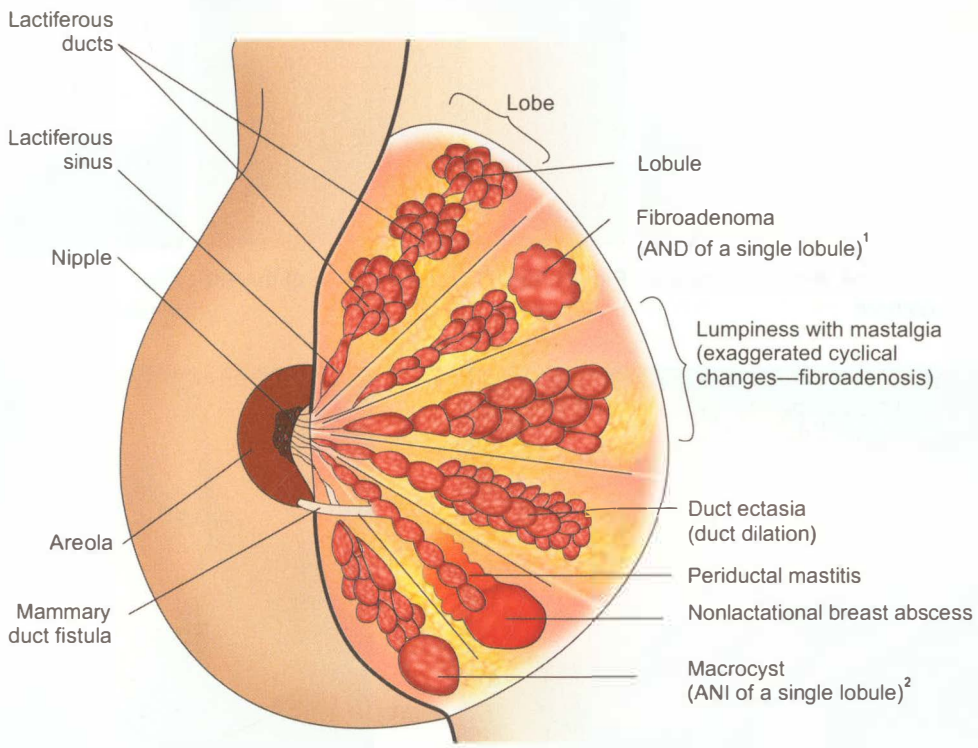


Fig. 21.19: Benign breast disorders and diseases (*Courtesy:* Dr Stanley Mathew, Professor of Surgery, KMC, Manipal)

[1]***Note**: Fibroadenoma is an **AND** (Aberration of Normal Development) of a lobule.*
[2]*Cyst (macrocyst) is an **ANI** (Aberration of Normal Involution) of a lobule.*

Table 21.1 Aberration of the Normal Development and Involution (ANDI) of the breast

Stage (Peak age in years)	Normal process	Aberration		Disease state
		Underlying condition	Clinical presentation	
Early reproductive period (15–25 years)	Lobule formation	Fibroadenoma	Discrete lump	Giant fibroadenoma (more than 5 cm).
	Stroma formation Nipple eversion	Juvenile hypertrophy Nipple	Excessive breast development	Multiple fibroadenomas Submammary abscess/ mammary fistula
Mature reproductive period (25–40 years)	Cyclical hormonal effects on glandular tissue and stroma	Exaggerated cyclical effects. Bloody discharge per nipple	Generalised or discrete lump	Cyclical mastalgia and nodularity (incapacitating)
Involution (35–55 years)	Lobular involution (including microcysts, apocrine changes, fibrosis, adenosis)	Macrocysts Sclerosing lesions adenosis	Discrete lumps X-ray abnormalities	Cystic diseases
	Ductal involution (including periductal round cell infiltrates), dilate with scleroma	Duct dilatation	Nipple discharge	Periductal mastitis with bacterial infection and nonlactational breast abscess—leading to mammary duct fistula
		Periductal fibrosis	Nipple retraction	
	Epithelial turnover	Mild epithelial hyperplasia	Histological report	Epithelial hyperplasia with atypia

***Terms to be avoided**: Fibroadenosis—replace with mastalgia and nodularity.*
Cystosarcoma phylloides—replace with phylloides tumour.
***Instructions to the students:** Carefully go through Tables 21.1, 21.2 and Fig. 21.1*
***References**: British Medical Bulletin: Volume 47, Number 2, April 1991.*

out malignancy by triple assessment (physical examination, mammography and cytology).

If it is not possible to conclusively rule out the possibility of malignancy, then the patient would be best advised a lumpectomy.

The term ANDI should not be confined to imply fibroadenosis (now termed mastalgia with nodularity). ANDI includes several aberrations and disorders (Table 21.1). The following table includes all the aberrations, disorders and disease entities originally included under the ANDI classification as proposed by LE Hughes, et al.

Breast pain

- It is for mastalgia that many women attend breast clinic. It can be classified as given in Table 21.3.
- If painful nodularity is present for more than one week, it is significant (more than normal discomfort).

CYCLICAL MASTALGIA WITH NODULARITY

Also called **mammary dysplasia**, fibrocystic disease, ***Schimmelbusch disease,*** hormonal mastopathy, or fibroadenosis.

Table 21.2 Classification of benign breast diseases

Nonproliferative disorders	Proliferative breast disorders	Atypical
• Cysts, apocrine metaplasia	• Sclerosing adenosis	• Atypical ductal hypoplasia (ADH)
• Duct ectasia	• Intraductal papilloma	• Atypical lobular hypoplasia (ALH)
• Calcification	• Ductal epithelial hyperplasia	
• Fibroadenoma and other related lesions	• Radial scar	
	• Complex sclerosing lesions	

Table 21.3 Mastalgia

Cyclical	Noncyclical
• Related to monthly cycles	• Less common
• Associated with premenstrual nodularity and breast discomfort	• Pain can be due to periductal mastitis or costochondritis (Tietz's disease)
• Excessive prolactin release from pituitary gland may be the cause	• Simple measures such as well supported brassieres
• Young women are affected	• Analgesics may be beneficial
• Reassurance	• Injection with local anaesthetic on a trigger point
• Drugs	
• Excision–last resort	

The term ANDI should not be confined to imply fibroadenosis (now termed mastalgia and nodularity). In fact fibroadenoma is an AND of a lobule and cyst (macrocyst) is an ANI of a lobule.

Definition

- It is an aberration of physiological changes that occur in the breast from menarche till menopause. It is an ANDI (aberration in normal development and involution).
- Women around the age of 40 are the usual sufferers.

Pathology (Key Box 21.7)

1. **Fibrosis** results in increased connective tissue growth. Fat and elastic tissue become less, and chronic inflammatory cells such as plasma cells, can be present.
2. **Cyst formation**: Fibrosis compresses the ductules, which is responsible for cyst formation. Hence, it is a retention cyst. The cyst contains dark mucoid material and it may discharge serous fluid or green coloured fluid through the nipple. Hence, it is called ***fibrocystic disease*** of the breast.

KEY BOX 21.7

PATHOLOGY

- Fibrosis
- Cyst formation
- Adenosis
- Epitheliosis
- Papillomatosis
- Apocrine metaplasia

Cyst may be single or multiple confined to one lobe or many lobes. These are microcysts.

3. **Adenosis:** Proliferation of the acini and gland is an important feature of fibroadenosis.
4. **Epitheliosis:** Fibroadenosis is not a precancerous condition but if the degree of **epitheliosis** is more, it can be considered as **premalignant** condition. Epithelial hyperplasia mainly occurs in the acini.
5. **Papillomatosis** and **apocrine metaplasia** of the epithelium lining cystic spaces are the other features.These changes are not considered premalignant.

Clinical features

- Females around the age of 30–40 are the victims—spinsters, married childless women and women who have not suckled their babies are the usual sufferers.
- Severe pain in the breast in the premenstrual period and during menstruation. **It is called cyclical mastalgia**. Upper outer quadrant, bilaterally is affected.
- Clinical examination of the breast reveals a coarse, nodular, tender, lumpiness which is better felt with the finger and the thumb. Often, there are multiple, irregular, firm, nodularities palpable bilaterally, especially in the upper outer quadrants. Nipple discharge which is serous or green coloured may occur.

Fig. 21.20: Submammary incision is given to excise a large lump in the breast

Fig. 21.21: Observe the nodularity of the lump

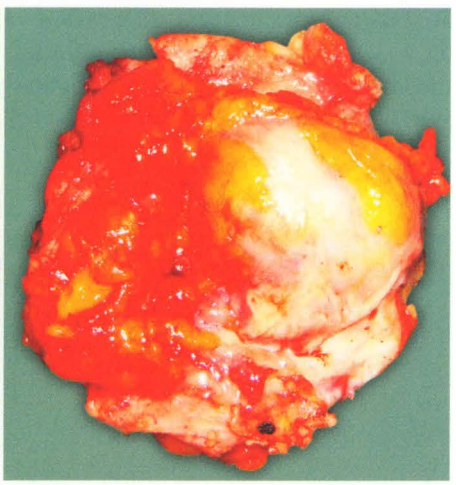


Fig. 21.22: Excised specimen having nodularity and cystic changes

Treatment (Key Box 21.8)

I. Conservative line of management

- **Evening primrose oil** in adequate doses for 3 to 4 months are beneficial in a few patients. These patients have abnormal fatty acid profiles and decreased levels of metabolites of linolenic acid. Treatment with primrose oil improves essential fatty acids because it is rich in polyunsaturated essential fatty acids (oleic acid and linoleic acid). Costly but still worth trying as first line of treatment, specially useful in women above the age of 40 (Key Box 21.9).

KEY BOX 21.8

TREATMENT

- Reassure
- Evening primrose oil
- Bromocriptine or danazol
- Tamoxifen
- Surgery

KEY BOX 21.9

EVENING PRIMROSE OIL

- First choice of treatment of cyclical mastalgia is evening primrose oil (EPO)
- Useful to treat mild to moderate cyclical mastalgia
- EPO contains 7% linolenic acid and 72% linoleic acid
- Dose is 1000 mg capsule—6 per day
- EPO can be taken with oral contraceptive pills
- No side-effects

- **Bromocriptine** which decreases the prolactin levels, 1.25 mg, twice a day, may reduce the pain and may be increased to 2.5 mg twice a day. It is useful for cyclical mastalgia.
- **Danazol,** it is a gonadotrophin releasing hormone inhibitor. 200–400 mg/day, thrice a day is given. It acts by reducing FSH and LH levels.
- **Tamoxifen** 10 mg, twice a day is a better alternative to danazol.

Treatment may have to be continued for some months (Table 21.4).

PEARLS OF WISDOM

Diuretics have no role. Topical NSAID application may help.

PEARLS OF WISDOM

Danazol is the best drug in cases of noncyclical mastalgia but it is teratogenic. Hence, the patient should take precautions against pregnancy.

II. Surgery

Indications for surgery in fibroadenosis:

1. FNAC suggests **epitheliosis**
2. A very **painful lump**
3. A hard lump about which the patient is worried and anxious
   - **Excision of the lump.** Surgery for fibroadenosis ends up in removing some breast tissue and the lump. As it has no capsule of its own, it is a messy surgery unlike fibroadenoma surgery (Figs 21.20 to 21.23).
   - Is there a role for **subcutaneous mastectomy?** In patients with family history of breast carcinoma, if they have

Table 21.4 Drug treatment of cyclical mastalgia

Drug	Mechanism of action	Dose	Maintenance dose	Side-effects
1. Evening primrose oil	• Contains essential fatty acids which correct abnormal prostaglandin synthesis. • Natural form of gamolenic acid	• 1000 mg/day 6 capsules • First choice in cyclical mastalgia • 4 months treatment	• Can be reduced to 3 capsules/day • Mild to moderate mastalgia	No side-effects
2. Danazol	Interfere with FSH and LH	• 50 mg/day—increased to 100 mg/day	• 50 mg/day for 3 months • Severe breast pain with nodularity	Amenorrhoea Weight gain Acne, hirsutism
3. Bromocriptine	It lowers prolactin by blocking its release from pituitary	• 2.5 mg/day—slowly increased to 2.5 mg twice/daily	• 2.5 mg/day • Inferior to danazol	Nausea, vomiting, dizziness
4. Tamoxifen	Antioestrogen	• 10 mg/day/3 for months	• Only for 3 months • Start if relapses occur	Minimal when used for short period.
5. Goserelin	Luteinising hormone Releasing hormone analogue	• 98% success in cyclical mastalgia	• Reserved for refractory cases	Reversible postmenopausal symptoms

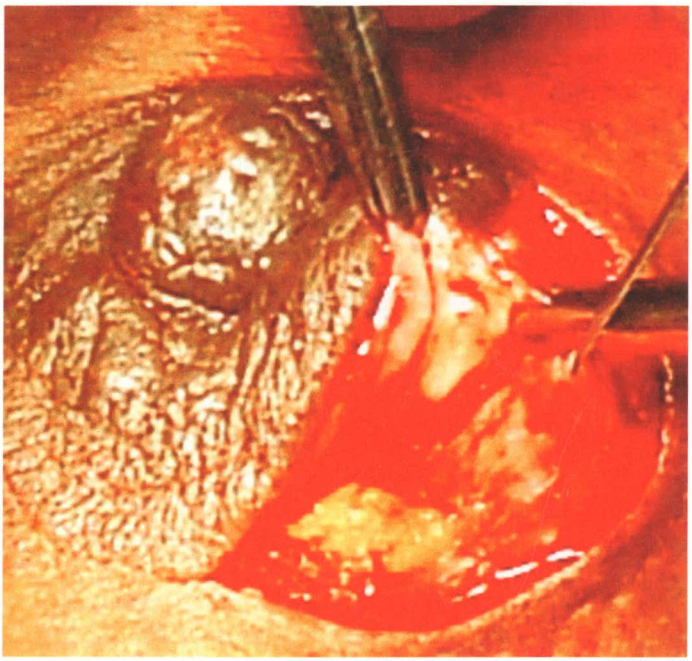


Fig. 21.23: A circumareolar incision is given to excise the lump

lumps in the breast with severe degree of epitheliosis, it may be worthwhile doing subcutaneous mastectomy.

FIBROADENOMA

It is a benign tumour in which the epithelial cells are arranged in a fibrous stroma. It is an **AND** (**A**berration of **N**ormal **D**evelopment) of a single lobule.

Types

1. Pericanalicular type in which fibrosis is more
2. Intracanalicular type in which fibrosis is less.

PEARLS OF WISDOM

- ***Small fibroadenomas (1 cm in size or less) are considered normal.***
- ***Larger fibroadenomas (up to 3 cm) are disorders.***
- ***Giant fibroadenomas (more than 3 cm) are a disease.***

Clinical features

- Peak age of incidence is at 20 years.
- Patients present with painless lump in the breast.
- It is smooth, round bordered, firm to hard in consistency, and freely mobile within the breast.
- Due to its free movement within breast tissue, it is known as **breast mouse**.
- However, when fibroadenoma occurs in elderly patients, it may not have characteristic features because of fibrosis.

CLINICAL NOTES

A paediatrician's mother, 52-year-old, underwent surgery for fibroadenosis. The pathology report was fibroadenosis with severe epithelial dysplasia. She came for follow up after 3 months. There were no fresh changes in the breast. However, after 9 months of surgery she presented with a hard lump with significant nodes in the axilla. She underwent Patey mastectomy. The case illustrates, epitheliosis is undoubtedly premalignant. Should the lady have been advised to undergo subcutaneous mastectomy after the first surgery?

Treatment

- Excision of the lump
  1. In **intracanalicular** type (Fig. 21.23), the lump is deeper and peripheral. It is removed by submammary incision.
  2. In **pericanalicular** type (Figs 21.24 and 21.25), the lump is superficial. It is removed by periareolar incision

Complications

- **Fibroadenoma and malignancy:** Patients with **simple** fibroadenoma and no family history of breast cancer have no risk of cancer. **Complex** fibroadenomas which show cysts, sclerosing adenosis, calcification have increased risk (3 to 4 times) of cancer.

Fibroadenoma and fibroadenosis are compared in Table 21.5.

DUCT ECTASIA / PERIDUCTAL MASTITIS PLASMA CELL MASTITIS

- Common in middle aged women.
- There is primary dilatation in one of the lactiferous ducts.

Aetiology

- Actual cause is not known. **Mild low-grade infection** (anaerobic bacteria) has been considered as one of the factors.
- Increased in **smokers**: Smoking increases the **virulence** of commensal bacteria.

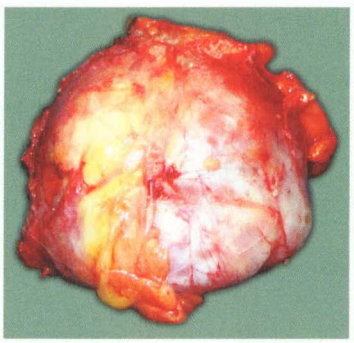


Fig. 21.24: Fibroadenoma excised—external view

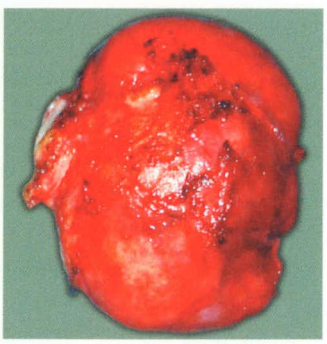


Fig. 21.25: Fibroadenoma—excised specimen

Table 21.5 Differences between fibroadenoma and fibroadenosis

	Fibroadenoma	Fibroadenosis
• Incidence	Less common	More common
• Nature	Benign tumour of one lobule	Aberration of normal changes in the breast
• Pain in the breast	Not a feature	Very common, premenstrual
• Location	Unilateral	Usually bilateral
• Lump	Well-defined, firm, mobile	Irregular, ill-defined, tender lump
• Capsule	Well-defined	No capsule
• Discharge	No	Serous or green coloured
• Malignancy	Rarely–sarcoma	Carcinoma (if epitheliosis is present)
• Treatment	Excision	Excision or drugs

Pathology

- There is dilatation of one of the lactiferous ducts.
- Due to dilatation, the contents tend to undergo stasis. The epithelial debris, and serous fluid collectively form a thick paste-like material rich in lipid.
- It may cause discharge per nipple
- There is **intense periductal inflammation with lymphocytes and plasma cells** (Fig. 21.26). Hence, it is called plasma cell mastitis (Key Box 21.10).
- Fibrosis causes nipple retraction

Clinical features

- Middle-aged woman
- **Paste-like material** discharge per nipple
- After some time, because of fibrosis, a lump can be felt, which can be confused for carcinoma of breast.
- **Bilateral slit-like retraction of nipple** of long duration
- Recurrent abscess and recurrent fistula are other complications.
- Routine mammogram—microcalcification
- Palpable subareolar mass

Management

- FNAC to confirm diagnosis.
- Antibiotics, drainage, excision of all major ducts (Fig. 21.27)

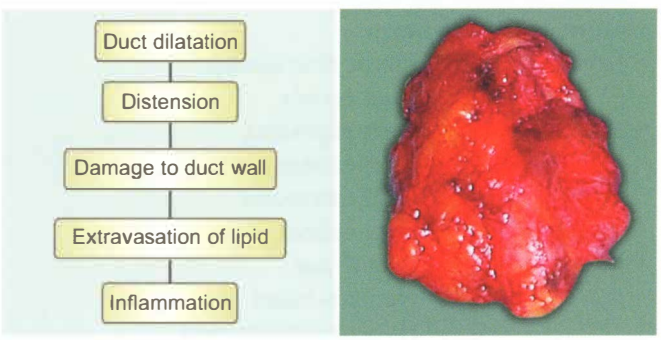


Fig. 21.26: Excised specimen. Observe the fibrosis due to periductal inflammation

KEY BOX 21.10

SUMMARY OF THE TREATMENT OF PERIDUCTAL MASTITIS

Periductal infiltration of plasma cells, lymphocytes
Excision of all major ducts (Hadfield's operation)
Retraction of nipple (partial)
Infection by anaerobes/irritation by smoking
Dilatation of breast /ducts
Untreated—Abscess/fistula/lump
Creamy/paste-like discharge
Treated by co-amoxiclav and metronidazole

Remember as **PERIDUCT**

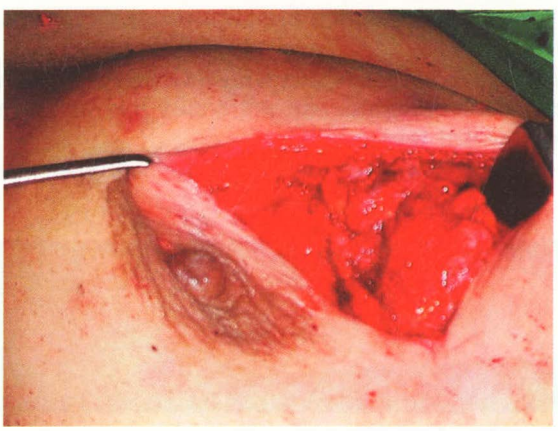


Fig. 21.27: Periductal mastitis at surgery. It was feeling hard (*Courtesy:* Dr Geetha, Associate Professor of Surgery, KMC, Manipal)

IDIOPATHIC GRANULOMATOUS MASTITIS

- Often patients present with features of mastitis (nonlactating) with pain, nipple retraction, lump in the breast.
- Some of them are treated with antituberculous treatment with FNAC, trucut or even biopsy findings of granuloma but they are nontuberculous.
- When tuberculosis, sarcoidosis, diabetes and Wegener's granulomatosis are excluded, the diagnosis of idiopathic granulomatous mastitis is made.

- Characteristic histological features include multinucleated giant cells, epitheloid histiocytes—noncaseating.
- Possible aetiological factors are trauma, hyper prolactinaemia, local irritation.

Treatment

- Symptomatic treatment to decrease pain and fever
- Steroids—30–40 mg prednisolone for 3 to 6 months period.
- Refractory cases need to be treated with surgery.

MACROCYSTS

- They occur due to excessive secretion of thin fluid within the lobules which enlarge to produce a cyst. Clinically, they present as firm swelling in the breast. It is an ANDI of a single lobule (firm because it is tense).
- They can be single or multiple. One of them can have huge dimension having a thin bluish capsule—***Blue domed cyst of Bloodgood*** (Figs 21.28 to 21.30 and Key Box 21.11).

KEY BOX 21.11

BREAST CYSTS

- Maximum incidence is between 40 and 50 years of age.
- Smooth and tense—hence feel firm on palpation
- One-third of patients may have more than one cyst in the breast.
- Diagnosis is by clinical examination
- If you suspect cyst, the investigation of choice is ultra-sonography.
- Increased incidence of benign breast lumps including cysts have been reported with prolonged use of **hormone replacement therapy.**

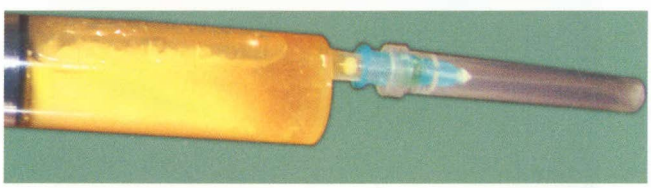


Fig. 21.28: Aspirated fluid—yellow colour in simple cyst

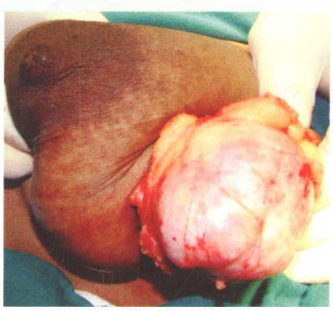


Fig. 21.29: Submammary incision—cyst delivered

Fig. 21.30: Fibrocystic disease—nature of fluid

(*Courtesy:* Dr Gabriel Rodrigues, Prof of Surgery, KMC, Manipal)

Treatment

Excision of cyst in the following situations:

1. Recurrence after 2 aspirations.
2. Blood stained aspirate.
3. Residual lump after aspiration.

GALACTOCOELE (Fig. 21.31)

- It is a solitary, subareolar retention cyst filled with milk. It occurs and dates back to the lactational period.
- It occurs due to inadequate drainage of the milk added by epithelial debris which block the lactiferous duct. Once the duct is blocked, proximally, the milky fluid accumulates resulting in a huge enlargement of breast.
- Rarely, they undergo calcification.
- It is treated by repeated aspirations. Duct patency can be restored by using 4-O nylon.
- Excision is the last choice.

Fig. 21.31: Galactocoele

DISCHARGE PER NIPPLE

Nipple discharge

- It is a common problem encountered in the outpatient department. It can be physiological such as lactational and or pathological from various causes.
- The clinical evaluation of the case of nipple discharge consists of following history examination.

1. Nature of the discharge

• Serous	Fibrocystic disease Duct ectasia
• Bloody	Duct papilloma Duct ectasia Duct carcinoma
• Greenish	Duct ectasia
• Milk	Lactational Nonlactational— Hyperprolactinaemia Oestrogen replacement therapy
• Yellow (purulent)	Breast abscess

2. **Unilateral or bilateral:** Duct papilloma is unilateral; fibrocystic disease and hyperprolactinaemia are bilateral.
3. **Single or multiple ducts:** Duct carcinoma/duct papilloma involves single duct, but fibrocystic disease affects multiple ducts.
4. **Spontaneous discharge** is typical of duct papilloma. Blood stained discharge on pressing the mass may be seen in carcinoma breast.
5. **Related to menstruation:** This is seen in fibrocystic disease and patients who are taking oestrogen replacement therapy.
6. **Discharge with mass:**
   Tender mass—fibrocystic disease breast abscess
   Nontender mass—carcinoma

Localisation

- Mammogram and ultrasound to detect any mass, any cystic lesions. Very often they do not help in the diagnosis.
- Prolactin levels, thyroid hormone profile (rarely hypothyroidism can cause discharge per nipple)
- Cytological examination of bloody discharge for malignancy
- Ductoscopy has been done but not very successful.

Treatment

- Rule out carcinoma—first
- Watery/serous discharge needs reassurance after ruling out fibrocystic disease
- A duct papilloma requires microdochectomy
- Discharge from multiple ducts or origin of the discharge is not clear—'core excision' of major ducts should be done (Hadfield operation).

GALACTORRHOEA

Secretion of milk unrelated to breastfeeding.

Causes

Physiological

- Stimulation during sexual activity.
- Extremes of reproductive age (puberty and menopause)

Drugs

- Oral contraceptive agents and oestrogen
- Antihypertensive agents such as methyldopa, atenelol, clonidine
- Dopamine receptor blocking agents such as chlorpromazine, haloperidol, metoclopramide.
- H_2 receptor antagonists such as ranitidine.

Pathological

- Pituitary adenoma/microadenoma.

Miscellaneous

- Ectopic prolactin secretion
- Hypothyroidism
- Chronic renal failure.

Treatment

- If the cause is identified, such as drugs mentioned above, stop them.
- If any other reason is detected, then treat the cause.
- Often no cause is identified, reassure the patient.

DUCT PAPILLOMA

- It is a benign lesion of the breast, usually single and unilateral, rarely multiple.
- Middle-aged women are affected and present with bleeding per nipple (Fig. 21.32).
- The tumour is situated in one of the larger lactiferous ducts.
- It presents as a small swelling just beneath the areola, palpation of which results in **discharge of blood**.
- Since, it is a *premalignant condition,* it is treated by microdochectomy.
- **Microdochectomy:** The opening of the lactiferous duct discharging blood is identified. It is probed with lacrimal probe or a straight needle. A small triangular piece of skin, along with the needle, and a wedge of breast tissue to a depth of about 4–5 cm is removed (Figs 21.33 and 21.34).

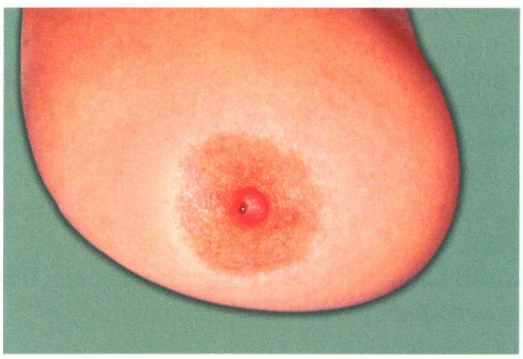


Fig. 21.32: Bleeding per nipple

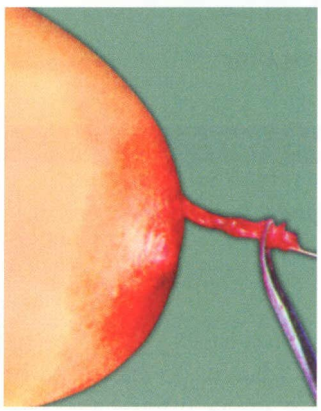


Fig. 21.33: Papilloma excision

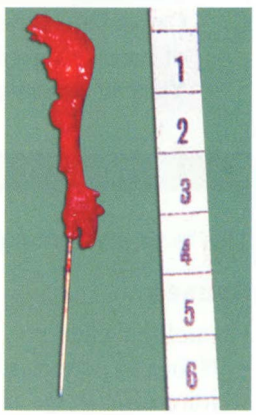


Fig. 21.34: Specimen of papilloma

- **Hadfield's operation:** Cone excision of the lactiferous ducts is done when duct of origin of nipple bleeding is uncertain. A circumareolar incision confined to one-third of the circumference is given and deepened. The flap is better not raised to avoid necrosis of areola. A core or cone of breast tissue from nipple to pectoral fascia or about 5 cm length is removed. **Most of the ductal papilloma will be located within 5 cm from the nipple.**

AXILLARY TAIL HYPERTROPHY

- More commonly it is the enlargement of axillary tail of Spence
- It is usually bilateral and feels like fatty tissue, lobular, soft to firm. This is how it is differentiated from lymph nodes in the axilla.
- Causes can be a physiological at puberty or during premenstrual/menstruation period or can be a part of 'ANDI'
- Pregnancy is also one of the causes of enlargement
- Rarely 'ectopic' axillary breast tissue other than tail of the breast may be present along the milk line.
- Usually reassurance is all that is required.
- *Pain* and *cosmesis* are the indications for removal.

TRAUMATIC FAT NECROSIS

T: Trauma either by a direct blow or by a seat belt or trivial contraction of pectoralis major. Sometimes no history of trauma.
R: Retraction of nipple, palpable hard lump, tethering of skin mimic carcinoma
A: Acids—fatty acids and glycerol released due to injury to the fat causes saponification
U: Unusually large, pendulous breasts are affected more often
M: Middle aged women with, microcalcification in mammography—mimics carcinoma
A: Age around 40 to 50 years
T: Treatment—Biopsy/excision of the lumps
I: Immediate reconstruction is possible
C: Can develop after tamoxifen therapy and, after any type of breast surgery
You can *remember as* **TRAUMATIC**

GYNAECOMASTIA

This is an *unphysiological enlargement of the male breast.*

Causes

Physiologic causes:
- Neonatal period, adolescence, senescence
- Idiopathic is the most common wherein no cause can be identified. However, there are many other causes of gynaecomastia which can be summarised as "**MASTIA**".
- **M**alignant tumour: Teratoma, bronchogenic carcinoma.
- **A**norchism: Absent testis
- **S**ex chromosome anomaly: Klinefelter's syndrome (XXY).
- **T**ablets: Cimetidine, stilboesterol, digitalis, spironolactone
- **I**diopathic: No cause is found
- **A**trophy of the testis: Liver cell failure, leprosy, etc.

Clinical classification of gynaecomastia

Grade I: Mild grade enlargement
Grade IIa Moderate grade enlargement
Grade IIb Moderate grade enlargement with skin redundancy
Grade III Marked grade enlargement with skin redundancy and ptosis (simulates a female breast)

Clinical features

- In idiopathic variety, gynaecomastia is bilateral. In other cases, it may be unilateral (Fig. 21.35).
- A "disk" like tender lump is palpable, with smooth surface.
- Examination should also include palpation of testis and to look for liver cell failure.

Treatment

Lumpectomy or mastectomy with preservation of nipple and areola (subcutaneous mastectomy).

Complications

- Rarely, gynaecomastia can predispose to male breast carcinoma (Fig. 21.36).

Fig. 21.35: Unilateral gynaecomastia—Often no cause is found: It is tender (*Courtesy:* Dr Surya Prakash Saxena, Chief Surgeon, District Hospital, Durg, Chhattisgarh)

Fig. 21.36: Gynaecomastia (*Courtesy:* Dr Satish Deshmukh and Dr Murtaza Akhthar, NKP Salve Institute of Medical Sciences, Nagpur)

CARCINOMA OF THE BREAST

Carcinoma of the breast is the major killer of middle-aged women in western countries. It is the second leading cause of cancer death. However, mortality from breast cancer is declining due to better screening and adjuvant treatment.

PEARLS OF WISDOM

However innocent looking the breast lump may be, it can be malignant unless proved otherwise.

Identifiable risk factors (Table 21.6 and Fig. 21.37)

1. **Women:** Women are 100 times more likely to have breast cancer as compared to men.
2. **Age:** Carcinoma breast is very rare below 20 years of age. More common in 35 to 75 years of age.
3. **Race:** Highest in whites, rare in Japanese and Taiwanese population
4. **Breast cancer and hereditary factors:**
   a. ***BRCA* I and *BRCA* II genes** have been found in long arm of chromosome 17 and 13 respectively in women with a family history of carcinoma of the breast. *BRCA* 1 and *BRCA* 2 are the genes associated with increased risk. ***BRCA* I and II mutations are more** common in **Ashkenazi Jews.** They are more prone for ovarian cancer also. Hence all patients with *BRCA* I and *BRCA* II mutations should consider a prophylactic bilateral oophorectomy after child bearing is completed, with bilateral mastectomy.
   b. **Cowden's disease** (multiple hamartoma syndrome)
   - Associated with reduced tumour suppressor gene PTEN
   - 30–50% of patients will develop breast cancer by 50 years of age.
   - The lesions found in this syndrome are **multiple facial trichilemmoma** (pathognomonic), oral papilloma, bilateral breast cancer, haemangiomas, lipomas, thyroid tumours, etc.

   c. **Ataxiatelangiectasia:** It is associated with haemangioma and carcinoma breast.

CLINICAL NOTES

A 35-year-old lady came to the hospital with fracture of the right femur. An intern, during history taking, elicited history of breast lump of 8 months duration. The patient reluctantly allowed the doctor to examine her breast as the lump was painless. She was found to have carcinoma breast with pathological fracture of femur.

   d. **Li–Fraumeni syndrome** is a rare disease with familial breast cancer and is associated with inherited mutation of tumour suppressor P53 gene.
   - It is a rare autosomal dominant disorder.
   - 90% of carriers will develop breast cancer by the age of 50.
   - They also can have other tumours in childhood such as soft tissue sarcoma, osteosarcoma, leukaemia.

PEARLS OF WISDOM

Breast cancer with BRCA 1 tends to be ER negative (ER–)
Breast cancer with BRCA 2 tends to be ER positive (ER+)

5. **History of breast cancer:** Risk of developing second breast cancer is about 0.5 to 0.7% in women with previous invasive breast cancers.

   Breast cancer is 3 to 4 times more likely to develop in women with a first degree relative who had breast cancer. This risk is further increased if they had premenopausal and bilateral cancer.

   Women with ductal carcinoma *in situ* (DCIS) are at an increased risk of developing ipsilateral and contralateral breast cancers (4.1% after 5 years).

6. **Benign breast disease:** In general **proliferative breast lesions are more vulnerable for malignancy.** Nonproliferative lesions such as cysts and duct ectasia do not increase the risk of breast cancer (Key Box 21.12).

Table 21.6 Lifetime cancer risk factors and relative risks

Relative risk < 2	Relative risk 2–4	Relative risk > 4
• Early menarche, late menopause	• Radiation exposure	• Radiation to mantle field in Hodgkin's lymphoma
• Nulliparous	• Breast cancer in one breast	• Ductal carcinoma *in situ*
• Age > 35 at first birth	• Mammographic dense breast	• Lobular carcinoma *in situ*
• Post-menopausal obesity	• One first degree relative (Mother/sister) with breast cancer	• Two first degree relatives with breast cancer
• Alcohol > 2 units/day		
• Hormone replacement therapy		
• Proliferative benign breast disease (LCIS and ADH)		

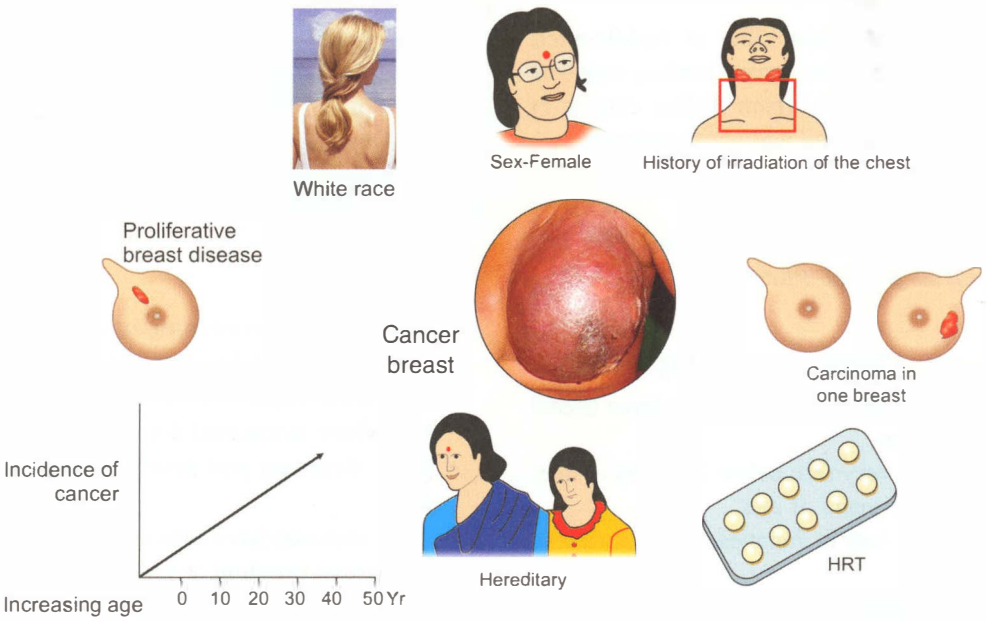


Fig. 21.37: Carcinoma breast—probable risk factors

KEY BOX 21.12

BENIGN BREAST DISEASE WITH RISK OF MALIGNANCIES

- Lobular carcinoma *in situ*/LCIS
  Ductal carcinoma *in situ*—DCIS—10 fold increase
- Atypical ductal hyperplasia—ADH/atypical lobular hyperplasia/ALH—5 fold increase
- Florid hyperplasia—2 fold increase
- Intraductal papilloma—2 fold incresae
- Radial scar and sclerosing adenosis: Both are microscopic findings. They can be felt as hard lesions. Radial scars are less than 1 cm. Doubtful role in malignancy or no increased risk.
- Fibroadenoma: No increased risk

7. **Diet:** Increased risk has been found in postmenopausal obese women and is due to increased synthesis of oestrogen (oestradiol) in the body fat. As a result of **aromatisation of androgens in adipose tissue, circulatory oestrogen levels are increased. Alcohol intake is associated with a 1.5 fold increased risk of breast cancer.** Vitamin C may have protective value. Increased intake of saturated fats and reduced intake of phyto-oestrogens increases risk.
8. **Endocrinal causes:**
   a. Longer the cumulative period of menstruation more the risk (early menarche and late menopause)
   b. More the cumulative period of lactation better the protection (more children, each child breastfed longer)
   c. More abortions and each occurring later increases risk.
   d. **Hormone replacement therapy (HRT)** in postmenopausal women increases incidence of breast cancer particularly when a combination of oestrogen and progesterone is used.
      - **Oral contraceptive pills are thought not to increase breast cancer.**
9. **Chest wall radiation:** Young children/women who have received mantle radiation for Hodgkin's disease have increased risk (19%) by the age of 50.
10. **Geographical:** Carcinoma of the breast is the disease of **white, western women. It is rare in Japan and Taiwan.** Genetic predisposition exists in a few cases, especially in bilateral breast carcinoma.

Pathology

- Majority of the tumours arise in the **ductal epithelium** (90%) which is called the ductal carcinoma and about 10% arise within **lobular epithelium** (lobular cancer).

DESCRIPTION OF THE TERMS USED PREVIOUSLY

Scirrhous carcinoma (Fig. 21.38)

- It is the most common form seen in about 60–75% of patients. In this variety, there is increased fibrotic reaction and less cellular reaction. It presents as a hard lump. Hence the name, scirrhous carcinoma. It produces grating sound when it is cut. Cut surfaces are concave. The chalky white necrosis and calcification may be visible occasionally. This

is the type of lesion which produces retraction of the nipple, infiltration of the skin and fixity to the chest wall.
- Malignant cells are round to polygonal with dark nuclei. Perivascular and perineural space infiltration is common.
- Atrophic scirrhous is an infiltrating duct carcinoma seen in elderly patients when there is atrophy of the breast.

Medullary carcinoma of the breast (Fig. 21.39)

- It is seen in around 15% of cases. It tends to occur in **well-formed breasts** in the reproductive age group and it feels more soft than hard. In addition to undifferentiated cells, occasionally well differentiated gland formation is present. Hence, the name, medullary adenocarcinoma. Presence of lymphatic infiltration is thought to represent a good host response, thus indicating a good prognosis.

Inflammatory carcinoma (Fig. 21.40)

- It constitutes less than 1% of all cases of carcinoma breast.
- Dermal lymphatic invasion is characteristic.
- Predominantly seen during pregnancy and lactation.
- Malignancy grows so rapidly that it invades more than half of the breast tissue. It comes under locally advanced breast cancer (LABC).
- Redness, pain and enlargement appear so suddenly that it is diagnosed as breast abscess. Hence the name, ***mastitis carcinomatosa***. It is differentiated from breast abscess by absence of fever and presence of gross *peau d'orange* due to blockage of subdermal lymphatics.
- Blunder biopsy (incision) should not be done thinking that it is an abscess (Fig. 21.41).
- This variety has the worst prognosis.
- High angiogenic and angioinvasive capability. Most of them are ER negative.

PEARLS OF WISDOM

Mastitis carcinomatosa comes under Stage T4D.

Paget's disease of the nipple (Fig. 21.42)

- It is a misnomer. It is ***not a disease of the nipple*** but an ***intraductal carcinoma*** involving excretory ducts that infiltrates nipple and areola early.
- Nipple can be ulcerated, fissured and cracked. Oozing is present.
- In advanced cases, entire nipple is destroyed (ulcerated).
- Lump appears much later than changes in the nipple.

Fig. 21.38: Scirrhous carcinoma—with nipple retraction

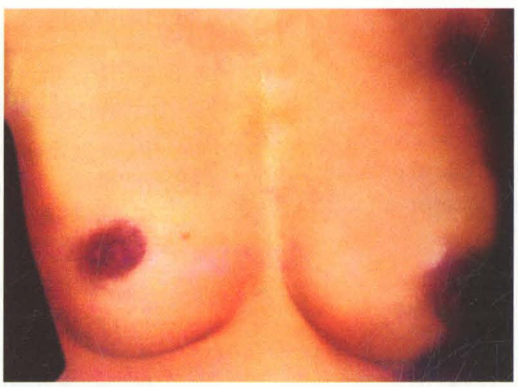


Fig. 21.39: Medullary carcinoma—common in middle age

Fig. 21.40: Inflammatory carcinoma—stage T4D

Fig. 21.41: Another case of inflammatory carcinoma. It has been incised mistaking it for an abscess

Fig. 21.42: Paget's disease of the nipple—totally destroyed nipple

- Microscopically, large **hyperchromatic cells** with clear cytoplasm or ***clear halo*** are seen.
- It is due to intracellular accumulation of **mucopolysaccharides**. These cells are called **Paget's cells**.
- It is differentiated from eczema as follows (Key Box 21.13).

KEY BOX 21.13

ECZEMA	PAGET'S DISEASE
• Seen in lactating women	• Seen in elderly women
• Bilateral	• Unilateral
• Itching is present	• Itching is absent
• Responds to antibiotics and local treatment	• Does not respond to antibiotics

Colloid carcinoma

- It is diagnosed because of production of mucin, intracellularly and extracellularly.
- Prognosis of this variety of carcinoma breast is better than other infiltrating duct carcinomas.

Definitions—current nomenclature

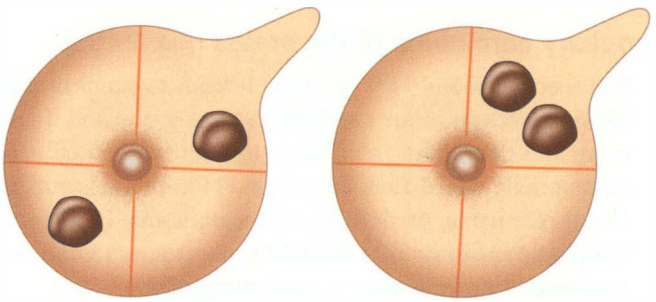
- **Carcinoma *in situ*:** This term is used when cancer cells do not invade through basement membrane. Examples: Lobular carcinoma *in situ* (LCIS) and ductal carcinoma *in situ* (DCIS) (Table 21.7).
- **Invasive carcinoma:** This term is used when cancer cells invade through basement membrane. A few examples are Paget's disease of the nipple, adenocarcinoma (scirrhous, simplex, medullary carcinoma, etc).

PEARLS OF WISDOM

Multicentricity refers to occurrence of a second in situ breast cancer outside the breast quadrant of primary in situ carcinoma.

Multifocality refers to occurrence of a second *in situ* breas cancer within same breast quadrant as the primary *in situ* cancer (Figs 21.43 and 21.44)

Upper and outer quadrant is commonly involved (60–65% because breast tissue is more there.



Figs 21.43 and 21.44: Multicentric and multifocal—this is the reason behind getting a mammogram before doing a breast conservative surgery

Invasive breast carcinoma

Depending upon specific histologic features, they are further classified into subtypes which are given below. However, it is interesting to note that 80% of invasive breast cancers are described as invasive, ductal carcinoma of 'no special type' (NST).

Foot-Stewart classification of invasive breast cancer

I. Paget's disease of the nipple
II. Invasive ductal carcinoma: 85% (Fig. 21.47)
   a. Adenocarcinomas [scirrhous, simple, NST (80%)]
   b. Medullary carcinoma 4%
   c. Mucinous (colloid) carcinoma 2%
   d. Papillary carcinoma 2%
   e. Tubular carcinoma 2%
III. Invasive lobular carcinoma 10%
IV. Rare types: Adenoid cystic, squamous cell, apocrine.

Table 21.7 Comparison of lobular carcinoma *in situ* (LCIS) with ductal carcinoma *in situ* (DCIS)

	LCIS	DCIS
• Origin	**Terminal Duct Lobular Units (TDLU)**	Epithelium that lines minor breast ducts
• Incidence in males	Not found	In 5% male breast cancer
• Multicentricity	60 to 90%	40 to 80%
• Bilateral occurrence	50 to 70%	10 to 20%
• Incidence of invasive cancer	25 to 35% of women	Increased by 5-fold
• Change into invasive cancer	LCI can give rise to both lobular and ductal invasive carcinoma	DCIS gives rise to invasive ductal cancer
• Age (years)	44–47	54–58
• Incidence	2–5%	5–10%
• Clinical sign	None	Mass, pain, nipple discharge
• Mammographic signs	None	Microcalcification

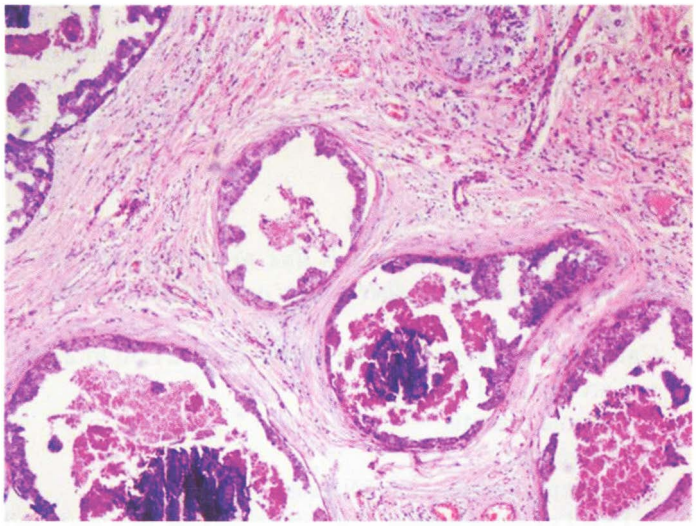


Fig. 21.45: Comedocarcinoma *in situ*—it is a DCIS—dilated terminal duct lobular units (TDLU) lined by malignant epithelial cells with luminal necrosis. No invasion into stroma. Slowly, the lumen gets occluded, can present as cyst and later calcification

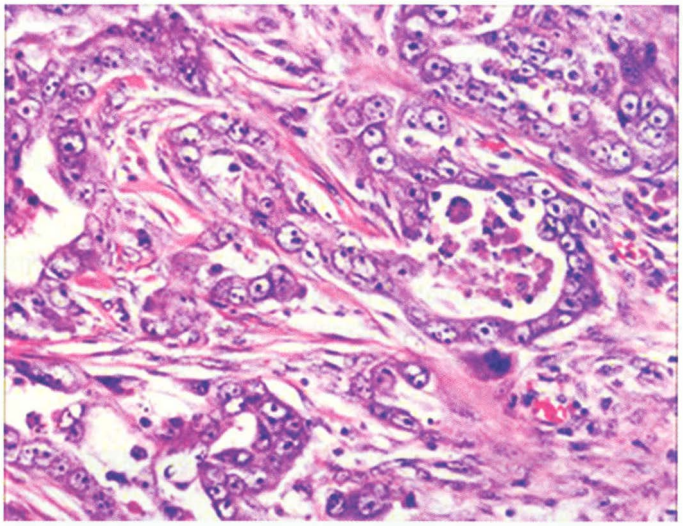


Fig. 21.47: Infiltrative ductal carcinoma: Infiltrating islands of malignant cells forming glands and trabeculae surrounded by desmoplastic stroma (*Courtesy:* Dr Laxmi Rao, HOD, Pathology)

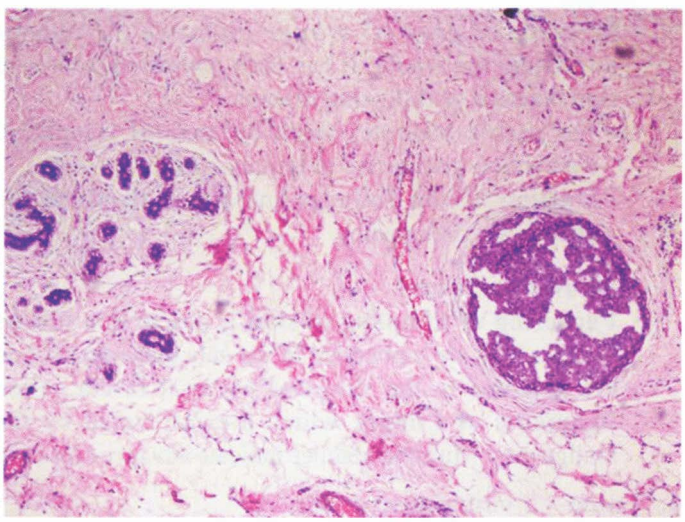


Fig. 21.46: Normal *vs* carcinoma *in situ*: Here you can see normal breast lobule (left) and *in situ* carcinoma (right) (*Courtesy:* Both Figs 21.45 and 21.46 by Professor Laxmi Rao, Head, Dept. of Pathology, KMC, Manipal)

NONINFILTRATIVE LESIONS

Carcinoma *in situ*

- It can be ductal—ductal carcinoma *in situ* (DCIS)
- Lobular—lobular carcinoma *in situ* (LCIS)

This is a type of **cancer without infiltrating** epithelial basement membrane. No lymph nodes will be enlarged. Hence, no role for axillary block dissection. 5-years disease free survival for DCIS is calculated by using modified Van Nuys prognostic index for ductal carcinoma in situ (DCIS).

PEARLS OF WISDOM

There is no role for chemotherapy in DCIS.

Modified Van Nuys Prognostic Index

Score	1	2	3
Size (mm)	≤ 15	16–40	≤ 41
Margins (mm)	> 10	1–9	> 1
Pathology	Non-high grade without necrosis	Non-high grade with necrosis	High grade with or without necrosis
Age (years)	> 61	40–60	≤ 39

5-year disease free survival for VNPI 4 to 6 = 100%.

PEARLS OF WISDOM

Lobular carcinoma is more dangerous because it is multifocal and bilateral.

- Lobular carcinoma refers to a lesion developing from the acini or terminal ductules of the lobule.
- More *in situ* carcinomas are diagnosed because of mammography and most of the breast conserving surgery is done in this group of patients.

Comedocarcinoma of breast (Figs 21.45 and 21.46)

- Comedos means cast or plug. It is a peripheral carcinoma wherein the tumour cells block the ductules by forming a case or plug producing a small cystic lesion. It is an example for intracystic carcinoma.

CLINICAL FEATURES OF CARCINOMA BREAST

- **Lump** in the breast is the most common presentation. It is the breast tissue in the upper and outer quadrant which is most frequently involved (65%). Typically it is hard and irregular but it can also be firm. Fixation to the skin, ulceration, *peau d'orange*, fixation to pectorals and chest wall occurs late (Figs 21.48 and 21.49).
- Bleeding per nipple is an uncommon symptom of carcinoma of the breast. **It involves multiple ducts and is unilateral**.
- About 5% of patients present with bony metastasis give rise to bony pains, i.e. pathological fractures, paraplegia, quadriplegia, etc.

Fig. 21.48: Scirrhous carcinoma presenting as hard lump with *peau d'orange*

Fig. 21.49: Carcinoma breast presenting as lump and ulceration

CLINICAL EXAMINATION OF A CASE OF CARCINOMA OF THE BREAST (Figs 21.50 to 21.59)

Inspection should be done in three positions:

I. Hands by the side of the patient

1. **Nipple**
   - **Bloody discharge** indicates duct carcinoma.
   - **Centrally retracted nipple**: Recent retraction indicate malignancy. It is due to fibrosis caused by extension o growth along the lactiferous duct. Remote retraction i either idiopathic or due to recurrent mild infectio (mastitis).

PEARLS OF WISDOM

Circumferential retraction is due to carcinoma. Slit-like retraction is due to duct ectasia or periductal mastitis.

Causes of retraction of the nipple (Figs 21.50 to 21.59)
- Carcinoma of the breast
- Chronic mastitis
- Congenital
- Chronic disease—tuberculosis
- **Level of the nipples:** The nipple may be elevated and retracted (Fig. 21.50) because of fibrosis induced by the growth. One should carefully watch for the level of nipple to detect an early case of carcinoma. Destruction of the nipple is a feature of Paget's disease of the nipple.

2. **Areola**
   - Presence of ***peau d' orange*** indicates the tumour infiltrating the areola. It has been compared to orange skin because of following reasons:
     - The areola becomes thick because of lymphatic obstruction giving rise to lymphoedema.
     - Fixation of hair follicles and sweat glands to the underlying malignancy.

3. **Skin over the breast**
   - **Puckering or dimpling of skin** is due to thin fibrous bands which are embedded in the subcutaneous fat and are attached to the skin and pectoral fascia called **ligaments of Cooper** which are infiltrated by the malignancy. Multiple nodules indicate advanced disease.

4. **Lump**—if visible, give details

5. **Oedema** of the arm is due to lymphatic blockage caused by lymph nodes in the axilla.

II. Hands raised above the head

1. ***Peau d'orange*** (on elevation of hands), becomes more prominent.

III. Bending forward

In cases of carcinoma infiltrating the chest wall, the breast will not fall forward on bending.

Palpation

1. **Local rise of temperature and tenderness** are usually not found in cases of carcinoma of the breast. However, rapidly growing carcinoma and inflammatory carcinomas do exhibit local rise of temperature, redness and tenderness.

Fig. 21.50: Slit like retraction

Fig. 21.51: Another case of nipple retraction—chronic abscess

Fig. 21.52: Retraction of the nipple—classical feature of intraductal carcinoma

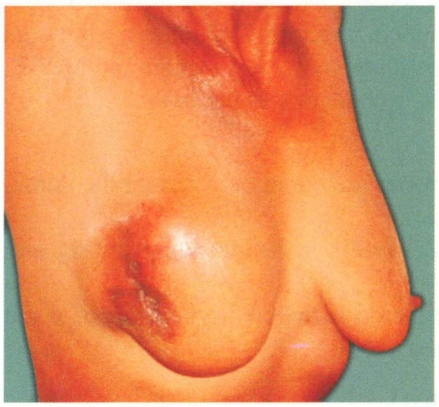


Fig. 21.53: Large lump with ulceration and bleeding

Fig. 21.54: Retracted nipple

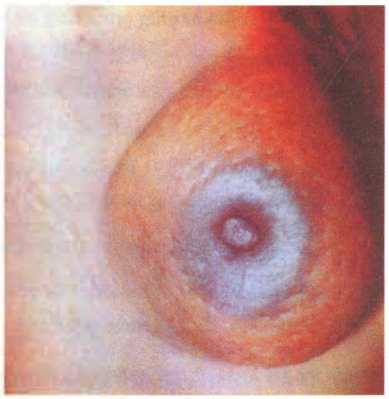


Fig. 21.55: *Peau d' orange*

Fig. 21. 56: Retraction of the nipple and *peau d' orange*

Fig. 21.57: Right nipple is elevated and retracted

Fig. 21.58: Paget's disease of the nipple (*Courtesy:* Dr Sreejayan, Prof. of Surgery, Calicut Medical College, Calicut, Kerala)

Fig. 21.59: Cancer en cuirasse (*Courtesy:* Dr Sreejayan, Prof. of Surgery, Calicut Medical College, Calicut, Kerala)

2. **Describe the lump:** The lump is the commonest presentation of carcinoma of the breast. The upper and outer quadrant is the commonest site of carcinoma of the breast because of more breast tissue in that quadrant. Typically, the lump is hard and irregular. However, very often carcinoma breast can present as a firm lump. In mastitis carcinomatosa the lump can be soft due to tumour necrosis.
3. **Intrinsic mobility** may be present but it moves with the breast tissue (fibroadenoma moves independent of breast tissue).
4. **Plane of the swelling**
   - Lift the skin. If it is not possible, it indicates that the tumour is fixed to skin.
   - ***Pectoralis major contraction test:*** Ask the patient to keep the hands on the flanks and press against the hip. If the lump cannot be moved after contraction, it indicates fixity to pectoralis major.
   - Fixity to the chest wall is assessed by 2 methods:
     1. A tumour which is fixed to the chest wall will not be mobile when pectoralis major is relaxed.
     2. Breast will not fall forwards.
   - ***Serratus anterior contraction*** test by pressing the hand against the wall. The test has to be done when the tumour is situated in the outer and inferior quadrant.

Axillary lymph nodes examination

- There are 5 groups of nodes in the axilla which are described under lymphatic drainage of the breast. However, very often, central group of nodes and pectoral nodes are enlarged. It is very difficult to feel the apical group of nodes.
- If the *axillary nodes* are *hard,* with or without *fixity,* they are significant.
- Soft to firm nodes need not be malignant but can be due to secondary infection because of fungating, ulcerating growth.

PEARLS OF WISDOM

Presence of supraclavicular lymph nodes does not indicate metastatic disease.

Examination for distant metastasis

1. Opposite axilla and opposite breast
2. Abdominal examination for secondaries in the liver which present as nodular liver, ascites and Krukenberg's tumour—bilateral bulky ovarian metastasis.
3. Rectal examination to rule out deposits in rectouterine pouch
4. Respiratory system examination to rule out effusion.
5. Bony tenderness should be looked for in the spine, long bones, skull, etc.

Spread of carcinoma of the breast

1. Local spread

As the tumour grows in size, it infiltrates the skin causing ulceration, fungation, bleeding, foul smelling discharge. Later, it involves pectoral muscles, chest wall, etc.

2. Lymphatic spread

- Central group, pectoral, lateral, subscapular and supraclavicular nodes.
- *Medial quadrant tumours* may involve the internal mammary glands in the upper three or four intercostal spaces, close to the sternum.
- Lymphatics from **inner medial quadrant** of the breast penetrate the rectus sheath and join the intraperitoneal lymphatics, thus producing ascites, Krukenberg's tumours (in premenopausal patients, ovary is vascular and fertile), rectovesical deposits, secondaries in the liver.

3. Blood spread

- Secondaries in flat bones are common (vertebral column, femur, ribs, scalp, etc).
- Secondaries in brain results in headache, vomiting and blurring of vision.
- Malignant pleural effusion is the common cause of death in carcinoma of breast.

Staging (*see* page 401)

INVESTIGATIONS

1. **Complete blood picture:** Hb% may be decreased.
2. Increased ALP levels in the blood suggests bone metastasis or liver metastasis
3. **Mammography** (Figs 21.60 to 21.63)
   - This has become the first diagnostic investigation
   - Diagnostic accuracy is about 90–95%.
   - It should always be combined with a clinical examination
   - Mammography is done when there is a doubt about the diagnosis

- Mammogram categories are grouped under—BI-RADS—Breast imaging reporting and data system

This is a classification system that is currently used in clinical radiology practice now.

Procedure

- A selenium coated X-ray plate is used directly in contact with the breast and the breast is exposed to low voltage and high amperage X-ray.
- 2 views: (a) Mediolateral and (b) craniocaudal

Indication

- Coarsely nodular breast
- Fibroadenosis
- Woman, aged 40 years with family history of breast cancer

TNM STAGING Breast cancer (Ca)

Tx	primary tumour cannot be assessed.
T0	no evidence of tumour
Tis	carcinoma in situ
Tis(DCIS)	ductal carcinoma in situ
Tis(LCIS)	lobular carcinoma in situ
Tis(Paget's)	Paget's disease of nipple with no tumour
T1	2 cm or less than in dimension
T1mic	microinvasion 0.1 cm or less in greatest dimension
T1a	0.1–0.5 cm in greatest dimension
T1b	0.5–1 cm in greatest dimension
T1c	1–2 cm in greatest dimension
T2	2–5 cm in greatest dimension
T3–	> 5 cm in greatest dimension
T4–	
T4a	extension into chest wall including intercostal muscles, ribs, serratus anterior (not pectoralis major)
T4b	extension into skin—peau d' orange, ulceration, cancer en cuirasse/satellite lesions (no puckering/ dimpling)
T4c	4a + 4b
T4d	inflammatory carcinoma/mastitis carcinomatosa
Nx	cannot be assessed
N0	no regional lymph nodes
N1	metastasis to mobile ipsilateral axillary lymph node
N2	metastasis to ipsilateral axillary lymph node
N2a	fixed/matted to one another or other structures
N2b	ipsilateral internal mammary nodes in the absence of clinically evident axillary lymph node
N3	
N3a	metastasis to ipsilateral infraclavicular lymph node
N3b	ipsilateral internal mammary and axillary lymph node
N3c	metastasis in ipsilateral suraclavicular lymph node
Mx	cannot be assessed
M0	no metastasis
M1	metastasis present

Regional lymph node – pN (pathologic)—done after axillary block dissection and i – immunohistochemistry

pNx	Cannot be assessed
pN0b	no regional lymph node metastasis histologically
pN0(i–)	no regional lymph node metastasis histologically, negative IHC
pN0(i+)	no regional lymph node metastasis histologically, positive IHC
pN0(mol–)	no regional lymph node metastasis, negative molecular findings (RT-PCR)
pN0(mol+)	no regional lymph node metastasis, positive molecular findings
pN1	metastasis in 1–3 axillary lymph nodes
pN1mi	micrometastasis (0.2 mm – 2.0 mm)
pN1a	1–3 axillary lymph nodes
pN1b	internal mammary nodes (not clinically apparent), microscopic disease in sentinel lymph nodes
pN1c	metastasis in 1–3 axillary lymph nodes and in internal mammary nodes (pN1a+pN1b)
pN2–	
pN2a	4–9 lymph nodes present
pN2b	internal mammary nodes + (clinically apparent)
pN3a	> 10 lymph nodes metastasis or metastasis to infraclavicular lymph nodes
pN3b	metastasis in clinically apparent internal mammary lymph nodes + positive axillary lymph node
pN3c	metastasis in ipsilateral supraclavicular lymph node

Instructions

- From undergraduate point of view, TNM classification is important
- What is given here is staging after an axillary block dissection. Hence, it is called pathological staging. If you know this, you may score higher marks.

STAGE GROUPING

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage IIa	T0	N1	M0
	T1	N1	M0
	T2	N0	M0
Stage IIb	T2	N1	M0
	T3	N0	M0
Stage IIIa	T0,T1,T2	N2	M0
	T3	N1, N2	M0
Stage IIIb	T4	N0–2	M0
Stage IIIc	Any T	N3	M0
Stage IV	Any T	Any N	M1

Advantages of mammography

- Noninvasive
- Minimum hazards of radiation

Disadvantages of mammography

- False positive cases are around 5%. Hence, mammographic positive cases should undergo FNAC to confirm the diagnosis.
- If a lesion is detected by mammography, but is impalpable, the following special techniques will help.

PEARLS OF WISDOM

In young women, microcalcifications are the only mammographic abnormalities suggesting malignancy.

Fig. 21.60: Architectural distortion

Fig. 21.61: Spiculation—medio lateral view—right side

Fig. 21.62: Spiculation—craniocaudal view—right side

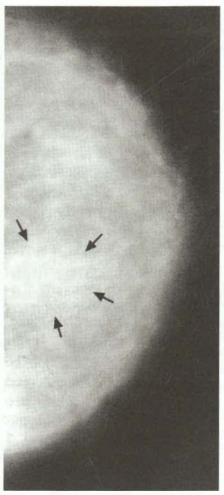


Fig. 21.63: Mammography showing a cystic lesion

BI-RADS scores range from 0 to 6

0: This score identifies a mammogram study that is still incomplete. The X-ray may have been cloudy, making it difficult to read the images. This can happen, for example, if the patient moved at the precise moment the picture was taken.

1: This score means that mammogram is negative (that is, no evident signs of cancer were found).

2: This score also means that mammogram is normal, with no apparent cancer, but that other findings (such as cysts) are described in the report.

3: A score of 3 means that mammogram is probably normal but **that there is an approximately 2 percent chance of cancer**. Hence, follow-up with a repeat mammogram in six months. And if there is a family or personal history of breast cancer, the radiologist may opt to do more tests at this stage rather than wait.

4: This score means that the findings on the mammogram are suspicious and that there is an approximately **20 to 35 percent** chance that a breast cancer is present. Core biopsy is a must in such cases for tissue sample.

5: This score means that the mammogram results are highly suspicious, **with a 95 percent** chance of breast cancer.

6: This means that patients have already been diagnosed with breast cancer and the pathologist has confirmed the diagnosis.

KEY BOX 21.14

MAMMOGRAPHY

- **Screening:** Asymptomatic women of more than 40 years.
- **Diagnostic:** Women with pain in the breast, mass, discharge, family history of breast cancer
- 2 views—Mediolateral oblique view is for outer quadrant and axilla. Craniocaudal view is for medial quadrants.
- Radiation dose is 0.1 centigray (cGy)—4 times that of chest X-ray dose but no side effects.
- Benign lesions are round, punctate, popcorn like, etc.
- Highly suspicious—pleomorphic, heterogeneous
  - Solid mass with **irregular edges, spiculation**
  - Long tentacles—tentaculation
  - Fine scattered calcification—**microcalcification**
  - **Distortion of architectural pattern** of the breast.
  - Asymmetrical thickening of breast tissues.

Technique of mammography

A. Double dye technique of injecting contrast medium and patent blue.

B. A hooked wire is pushed down the needle left in the breast tissue which acts as a guide to the surgeon—mammographic needle localisation and needle biopsy. For mammography findings *see* Key Box 21.14.

C. Biopsy from nonpalpable lesions: First mammography is done which may show suspicious areas such as microcalcification. Then ultrasound or stereotactic localisation is done, followed by FNAC. This is almost 100% accurate in the diagnosis of breast cancer (see under image guided biopsy).

PEARLS OF WISDOM

45% of breast cancers can be seen on mammography before they are palpable.

4. **FNAC** (Fine Needle Aspiration Cytology) is quick, safe and easy method in which a cytological diagnosis can be made. The accuracy is more than 95% with an experienced cytologist.
   - False negative 15% (infiltrating lobular carcinoma, tubular carcinomas may mimic fibroadenoma)
   - False positive 1% (atypical ductal hypoplasia, papilloma, radiation changes)
5. **Trucut biopsy:** If FNAC is negative, a trucut biopsy or **vacuum-assisted biopsy** (VAB) using 11 gauge biopsy probe can be taken. The advantage of these biopsies is

preoperative assessment of hormone receptors can be done. False negative (25%) and no false positive (0%).

6. **Incisional biopsy:** Under local anaesthesia, a small incision is made over the lump and a biopsy is taken, which is sent for frozen section. If frozen section report is malignant, an appropriate mastectomy is done. If the frozen section is negative, a lumpectomy is done and if report comes later as malignant, a mastectomy is done.

7. **Image guided biopsy in indeterminate lesions**
   a. **Ultrasound guided biopsy:** Microcalcifications are seen infrequently but if found, they can be biopsied using ultrasound as a guide. Success rate of positive histopathology is high (Fig. 21.64)
   b. **Wire localisation** (Figs 21.65 to 21.67)
      - **Indication:** Localising nonpalpable breast lesions.
      - **Procedure:** Mammogram is done and the suspicious area is localised (microcalcification). A hooked wire enclosed within a needle is placed within the breast under guidance. The needle is removed. Patient is shifted to OT. In the OT incision and dissection follows the wire till 'Hook' is reached and tissue is excised and sent for histopathology.
   c. **Stereotactic biopsy:** It is used to biopsy mammographically—suspicious lesions. Stereotactic biopsy techniques include—vacuum-assisted core biopsy.
   d. **Mammotome:** It is a hand held vacuum-assisted biopsy instrument which can be used under image (ultrasound or stereotactic) guidance. It gives accurate diagnosis of nonpalpable mammographic abnormalities.

8. **Chest X-ray:** Rule out pulmonary secondaries, effusion, or mediastinal widening (**2 types**) (Fgis 21.68 and 21.69)
   - **Cannon ball type** (haematogenous)—presents late
   - **Lymphangiectatic type** (lymphatic spread))—presents with intractable cough.

9. **Abdominal ultrasonography** is done to rule out secondaries in the liver, ascites, rectouterine deposits.
   - Incidence of liver metastasis in Ca breast is 6%. Routine USG abdomen is indicated only if hepatomegaly is present or any abnormality in LFT or in stage III or IV.

10. **Bone scan:** Incidence of bone metastasis (Fig. 21.70).
    - T1, T2—< 2%, no role for scan
    - T3, T4—25%. Even then, 60% of skeletal tissue should be demineralised before it is evident. Hence, bone scan is indicated if symptomatic, increased ALP or stage II and above.

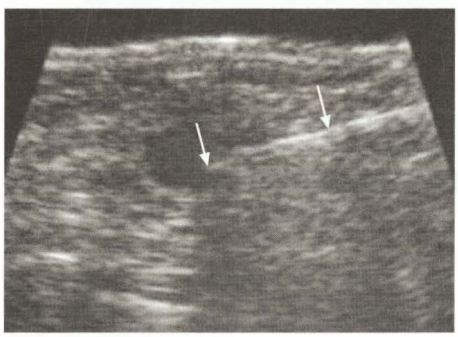
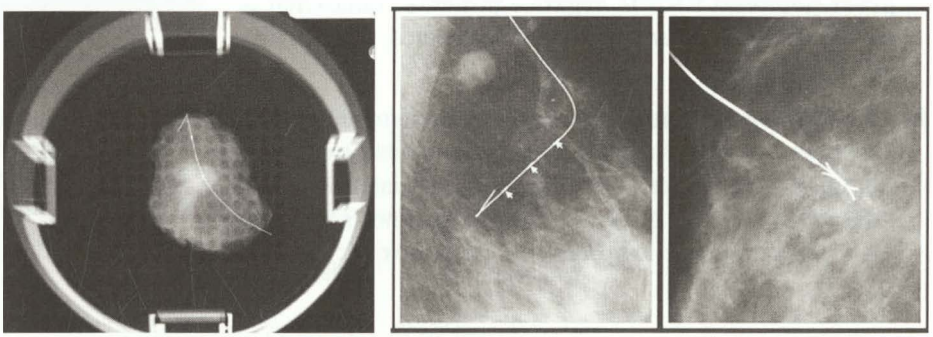


Fig. 21.64: Ultrasound detecting a lump in the breast and a wire is being introduced within



Figs 21.65 to 21.67: Mammography directed wire localisation (*Courtesy:* Dr Sampath Kumar, Professor of Surgery, Dr. Chandrakant Shetty, Professor of Radiology and Imaging, Dr Poornima, Resident, Dept of Surgery, KMC, Manipal)

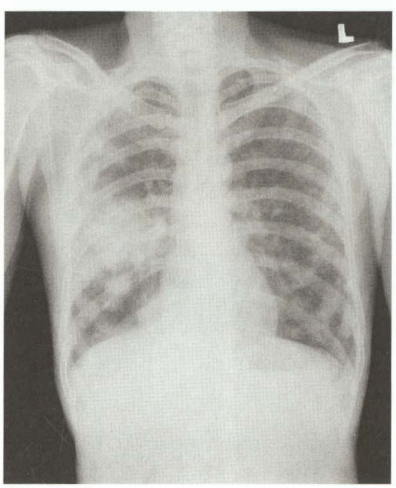


Fig. 21.68: Cannon ball secondaries

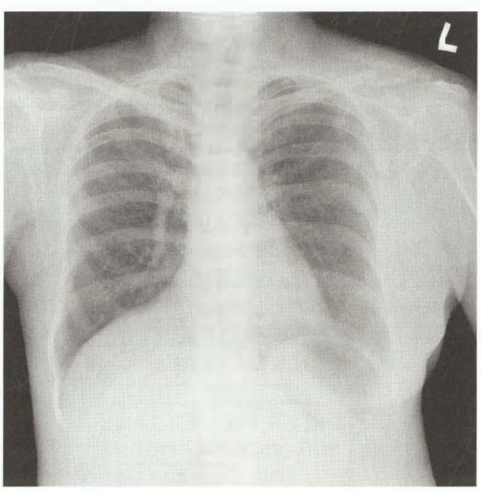


Fig. 21.69: Carcinoma breast with metastasis in the clavicle

Fig. 21.70: Bone scan Ca breast

11. Steroid hormone receptors—immunohistochemistry

A. Intracellular steroid hormone receptor proteins, primarily ER (oestrogen receptors) and PR (progesterone receptors) have shown to be indicators of prognosis and a guide to hormone and endocrine therapy. About 50 to 85% of invasive breast cancers contain measurable amounts of ER. The **concentration of ER increases with age** with both having their highest levels in postmenopausal patients.

- Normal value of ER is < 10 fmol/mg proteins. It is **considered positive** > 10 fmol/mg proteins. Upper levels as high as 1000 fmol/mg proteins may be there.
- The presence of ER implies that normal cellular mechanism for processing oestrogen has been maintained despite the malignant change. **Patients with ER positive tumours have longer disease-free survival** after primary treatment, superior overall survival and longer survival after recurrence compared with patients with ER-negative tumours.
- Before starting hormonal therapy steroid hormone receptors have to be checked and measured. Depending upon the levels, whether positive or negative, the hormonal therapy is given (details below).
- **All patients may benefit from tamoxifen except premenopausal ER/PR negative patients.** Tamoxifen is started only after chemotherapy is completed. It should be given for 5 years.

B. HER-2/neu receptor: It is a membrane tyrosine kinase receptor and a marker of cellular proliferation, expressed in up to 50% of cases. It is usually associated with ER negativity and high grade tumour, poor prognosis but there is a better response to adriamycin (Key Box 21.15).

KEY BOX 21.15

HER-2

- Measured by immunohistochemistry
- Test for gene amplification—fluorescent *in situ* hybridisation (**FISH**) is gold standard.
- 25% of all breast cancers overexpress HER-2
- It is a transmembrane tyrosine kinase receptor
- High grade tumours usually express this
- ER negative—poor prognosis
- Responds to 'TRASTUZUMAB'

Thus 'FISH' is done only when HER-2/neu is 2+

Interpretation of HER-2/neu result interpretation by IHC

1 + means negative
2 + means equivocal result—to confirm by 'FISH'
3 + positive or over expressed/amplified

C. Detailed histopathological examination and followed by immunohistochemistry (IHC) and DNA microarrays form an important part of treatment or the breast cancer. The new marker for cellular proliferation has been identified and named Ki 67 (Key Box 21.16).

KEY BOX 21.16

Ki 67

- Also known as MK167
- It is a marker for cellular proliferation
- It depicts growth fraction of all proliferation
- Higher values suggest aggressive tumour
- A cut off value of < 14% is generally used to denote low and high values

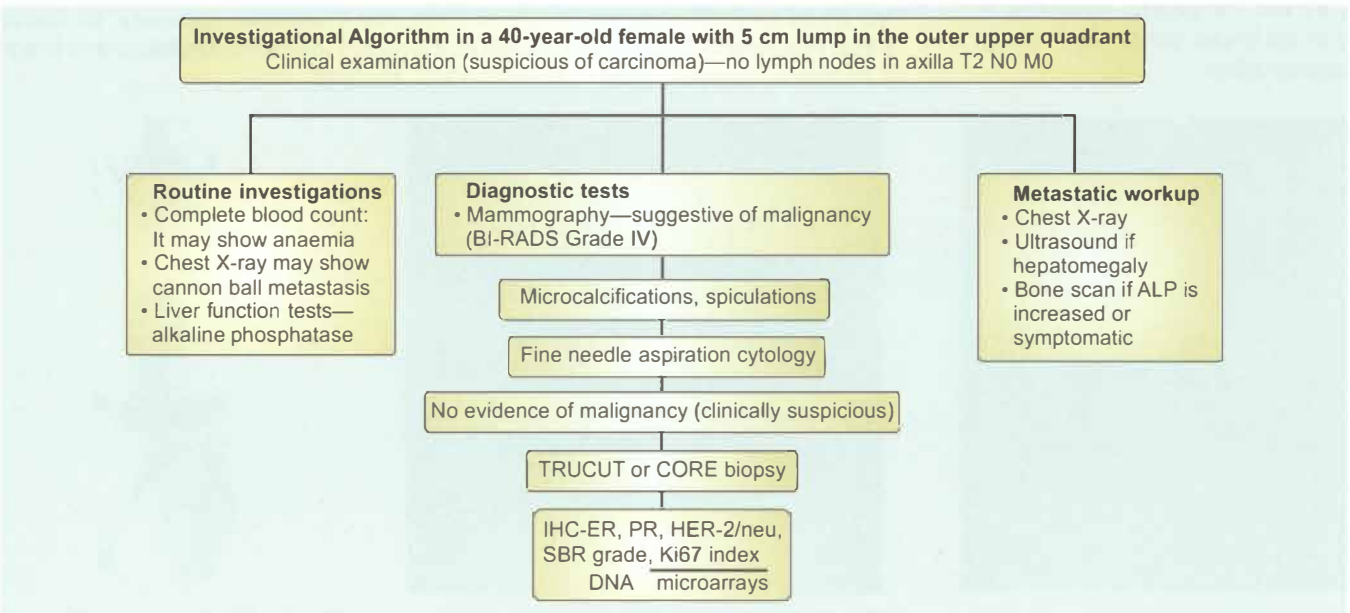


Fig. 21.71: Investigations in carcinoma breast

D. DNA microarrays: They compare DNA from normal cells to breast cancer cells. They are very expensive.

Examples:
1. Oncotype Dx—it is 21 gene assay
2. Mammaprint—70 gene assay

Based on these, treatment is divided into specific groups referred to as 'luminals'—this is the latest treatment and is given in page 422.

Pathological evaluation of carcinoma breast

- Tumour, node, metastasis (TNM), oestrogen receptor (ER)
- Progesterone receptor (PgR)
- HER—2, Ki 67 index.
- Tumour grade: **Scarff-Bloom-Richardson** (SBR).

SBR grade

Tumour grade: Depending upon three factors such as nuclear pleomorphism, tubule formation and mitotic rate, tumours are given scores and graded. Grade III tumours means they are poorly differentiated. Grading system is popularly called 'SBR' system—Scarff Bloom Richardson—Elston-Ellis modified 'SBR'grading system.

Nuclear pleomorphism:
Score 1—small uniform nucleoli
Score 2—intermediate nucleoli
Score 3—large prominent nucleoli

Mitotic count:
Score 1—< 10% mitosis in 10 HPF
Score 2—10%–20% mitosis
Score 3—> 20% mitosis

Tubule formation
Score 1— > 75% cells in tubule form
Score 2— > 10–75% cells in tubule form
Score 3—< 10% cells in tubule form

Grade	Score
Favourable	1
Unfavourable	up to 3
Grade I	3–5
Grade II	6–7
Grade III	8–9

Examples

Well-differentiated:	Grades	3, 4, 5
Moderately differentiated:	Grades	6, 7
Poorly differentiated:	Grades	8–9

12. Role of ultrasonography in breast lump

- It is second only to mammography.
- Define cystic lesions
- Demonstrate echogenic qualities of solid abnormality
- Well-circumscribed lesions are cysts with smooth margins (benign)
- Benign lesions are round to oval-shaped with smooth contours.
- Cancers have irregular walls.
- It can guide FNAC, core biopsy, etc.
- Painless, cost effective, can be reproducible but operator dependent.
- Not ideal for lesions which are 1 cm or less in diameter.

13. Role of MRI[1]

- Particularly useful in detecting malignancy when mammographically subtle or occult (lobular carcinoma).
- It can differentiate scar tissue from cancer. Hence can detect local recurrence after surgery.
- MRI is better than mammogram in assessing the response of the tumour to neoadjuvant chemotherapy.
- It is also better investigation in dense breasts and in pregnancy.
- Also for imaging of the breast with implants.

Summary of investigations in Ca breast (Fig. 21.71)

PEARLS OF WISDOM

When axillary nodes are positive for adenocarcinoma, but the primary is unknown, MRI is the ideal investigation to detect an impalpable breast lump.

14. Bone marrow aspiration

in cases of unexplained cytopaenia or leukoerythroblastic blood smear.

PEARLS OF WISDOM

Thus 'TRIPLE ASSESSMENT' is the most important method for the diagnosis of carcinoma. It includes:
- ***History and clinical examination***
- ***Imaging: Below 40 years—Ultrasound;***
  Above 40 years—Mammogram
- ***Fine needle aspiration cytology.***

Students are hereby instructed to follow the investigation which are available and routinely done in your hospital from the clinical point of view and select the investigation based on staging of the disease.

TUMOUR BIOLOGY

- Halsted did the first radical mastectomy for carcinoma breast in 1878. This was the gold standard operation for carcinoma breast for nearly 100 years. Carcinoma breast is considered to be a systemic disease to begin with according to **Fischer's theory** if we consider the biology of tumour growth.

[1]From practical point of view, the clinician should select the best investigation in each patient and not all the investigations given here.

Cell population and size of the tumour

10^1	< 0.5 cm	No independent blood supply, less aggressive
10^5	0.5 cm	Has its own blood supply
10^9	1 cm	Has its own blood supply

Hence tumours with 0.5–1 cm or more may exhibit haematogenous spread owing to their own blood supply. Therefore, it is logical to treat it as a systemic disease and apply some mode of systemic therapy for tumours > 1 cm.

Also, the other modalities of treatment such as radiotherapy, chemotherapy, hormonal therapy are playing a major role together with surgery. Hence, the current approach is to do minimal local surgery and aggressive approach towards management of lymph nodes or metastasis.

Stage wise carcinoma breast is divided into

a. Early breast cancer—Stages I and IIA
b. Locally advanced—Stages IIB and III
c. Advanced/metastatic—Stage IV

Surgical management is thus based on the stage of the disease. There is no confusion regarding management of either early breast cancer or metastatic breast cancer (MBC). **Controversy exists in the management of LABC (III) which has both operable and inoperable diseases which is given in page 411.**

TREATMENT OF EARLY BREAST CANCER—STAGES I, IIA

A. Primary: Growth and Axilla

- Locoregional control of primary disease is by **surgery** in the form of **modified radical mastectomy** (MRM) or breast conservative surgery (BCS).

B. Adjuvant

- Locoregional control of **residual disease** is by radiotherapy
- Systemic control of disease is by hormonal therapy or chemotherapy.

Surgery (Table 21.8)

1. **Wide local excision (lumpectomy)** is indicated in tumours less than 4 cm in size and with well-differentiated histology. It includes removal of the tumour with a rim of at least 1 cm of normal breast tissue. If the nodes are palpable and enlarged, this is combined with axillary block dissection, using separate incision. Currently, this procedure has become more popular. It is also called **Breast Conservative Surgery** (Key Box 21.17*). Contraindications for BCS are given in* Key Box 21.18.

KEY BOX 21.17

SALIENT FEATURES OF BCS

- Tumour size—4 cm or less
- Should be able to get 1 cm negative margin
- Availability of frozen section is desirable
- Should not do undermining of the flaps as we do in MRM
- Absolute haemostasis
- No suction drain to be kept
- Direct approximation of the skin with no attempt to close the fatty tissue.
- Decision about cosmetic acceptance to be done by patient specially to 4 cm

KEY BOX 21.18

CONTRAINDICATIONS FOR LUMPECTOMY (BCS)

- Multicentric disease (rule out by mammogram)
- Pregnancy
- Central quadrant tumour (relative)
- Prior radiotherapy to the breast
- Prior chest irradiation
- Collagen vascular diseases
- Recurrence after BCS
- Skin fixation, involvement of nipple, chest wall

Table 21.8 Different types of surgery for carcinoma breast

Types of surgery	What does it mean?	Comments
• Local wide excision	• Tumour along with 1 cm of normal tissue is removed with an ellipse of the skin over lump	• Most popular breast conservative surgery done in T1 and T2 cases
• Quadrantectomy	• Quadrant containing the tumour is removed	• Not done
• Total mastectomy and axillary clearance	• Entire breast tissue is removed. Both pectoralis minor and major are preserved	• Frequently done—good retraction of pectoralis major and minor are necessary for the axillary clearance
• Patey mastectomy	• Breast tissue + pectoralis minor is removed and axillary block dissection	• Mostly done and removal of pectoralis minor helps the axillary dissection
• Halsted radical mastectomy	• Entire breast tissue and both the pectoral muscles are removed.	• Not necessary as carcinoma breast is considered as systemic disease today.

TEN COMMANDMENTS

A few important tips about local wide excision—lumpectomy (Figs 21.72 and 21.73)

1. Incision is made directly over the tumour
2. Pectoral fascia is usually not opened unless the tumour is adherent
3. Perfect haemostasis
4. Tumour removed with 1 cm margin all around
5. Ideally specimen mammogram and frozen section is done to look for clearance
6. Cavity should not be closed or obliterated by sutures as small seromas get absorbed
7. For better orientation of the tumour a long silk is tied on the lateral side. Remember a **long lateral, short suture posteriorly.**
8. Metal clips or coloured beads can be applied.
9. Drain should not be kept
10. Anterior and posterior margins are less important if a full thickness of breast tissue is removed.

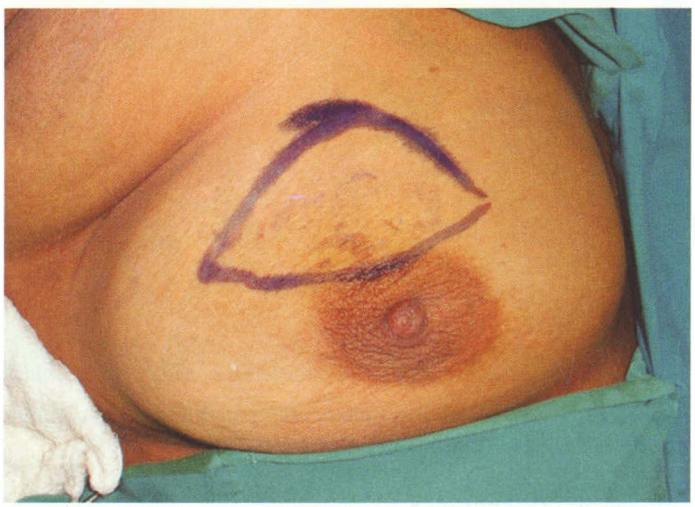


Fig. 21.72: Local wide excision is done—if the skin is involved, it is also removed, undermining of the flaps is not required

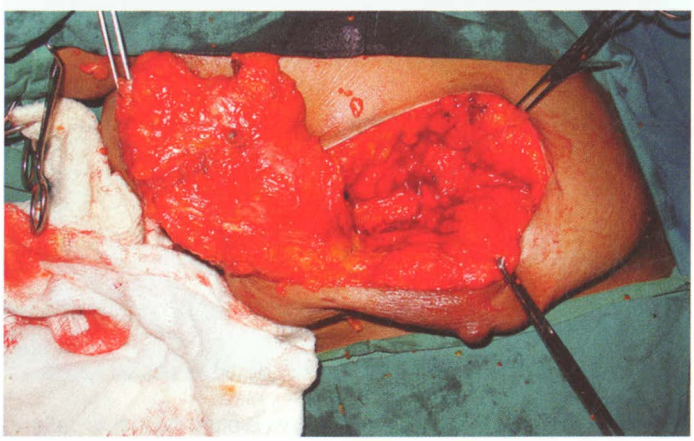


Fig. 21.73: Local wide excision is in progress. 1 cm of normal breast tissue of the cancerous lesion is all that is necesary

PEARLS OF WISDOM

Radiotherapy to the breast tissue is mandatory in all cases of breast conservation surgery.

2. **Total mastectomy with axillary clearance (MRM)** is equally good (good retraction of pectoralis minor facilitates axillary dissection—**Auchincloss** modification)
3. **Patey mastectomy:** This is the most acceptable and most widely practised surgery. It is also called ***modified radical mastectomy.*** In this, the entire breast including nipple and areola (simple mastectomy) are removed with, pectoralis minor, followed by axillary block dissection. A complete axillary block dissection should include node clearance up to level III (Figs 21. 74 to 21.79).
   - **Level I** : Extends from axillary tail to the lateral border of pectoralis minor.
   - **Level II** : Extends from lateral border of pectoralis minor to medial border of pectoralis minor.
   - **Level III** : Up to the apex of axilla.
4. **QUART therapy by Veronesi:** It includes **Q**uadrantectomy (the entire segment of the breast containing tumour is removed), **A**xillary block dissection and **R**adiotherapy to the breast or axilla.
   - However, quadrantectomy, by removing large amount of breast tissue, gives rise to poorer cosmetic results. It seems unnecessary to remove entire segment of the breast, when in reality breast cannot be strictly divided into quadrants (unlike hepatectomy). Hence, it is not very popular.

Advantages of Patey mastectomy over Halsted radical mastectomy

- Cosmetically better accepted as axillary fold is maintained.
- Function of the shoulder is better, and it gives a stronger and more useful arm.

5. **Radical mastectomy:** In this operation, following structures are removed
   - Entire breast including nipple and areola, skin overlying the tumour along with fat, fascia and lymphatics.
   - Axillary block dissection, including complete clearance of axillary fat and up to Level III nodes clearance.
   - Sternocostal portion of pectoralis major, entire pectoralis minor, few fibres and aponeurosis of internal oblique, serratus anterior, latissimus dorsi and subscapularis.

Three important structures should be preserved

- **A**xillary vein
- **B**ell's nerve (long thoracic nerve which supplies serratus anterior)
- **C**ephalic vein.

Disadvantages of radical mastectomy

- Mutilating surgery
- Poor cosmetic results
- Lymphoedema of arm
- High morbidity rate

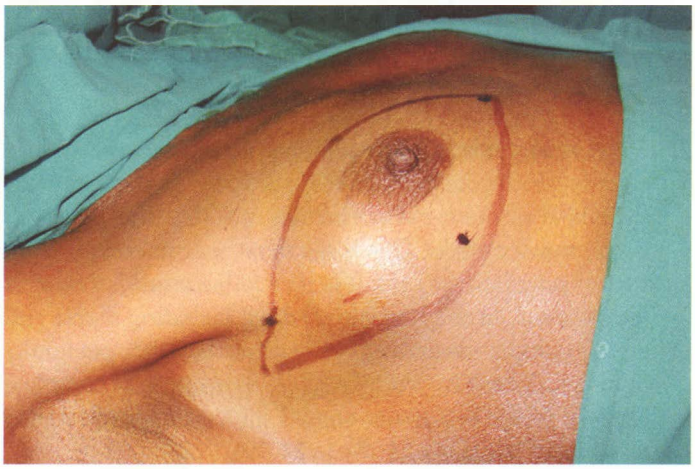


Fig 21.74: Classical MRM incision which includes nipple areola complex and slightly extending into axilla to facilitate axillary block dissection

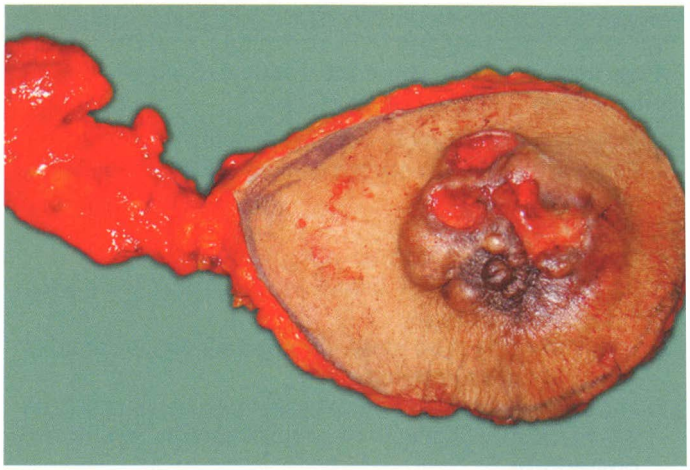


Fig. 21.75: Patey mastectomy specimen: In this operation, entire breast including axillary tail with all the axillary group of lymph nodes and pectoralis minor are removed

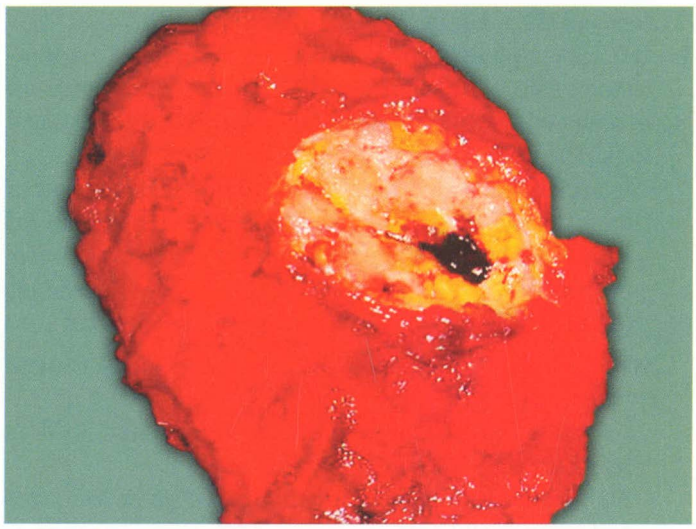


Fig. 21.76: Tumour with degeneration

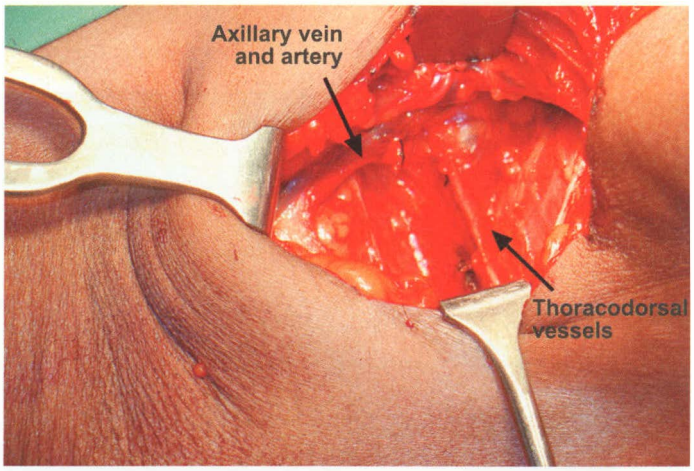


Fig. 21.77: You can see the axillary vein and thoracodorsal nerve as the axillary dissection is in progress

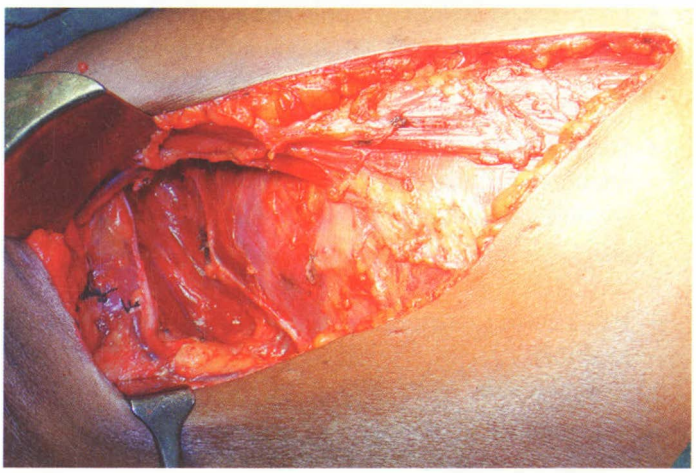


Fig 21.78: Same patient as shown in Fig. 21.77. Axillary block dissection is complete

Fig 21.79: Initially local wide excision was planned in this patient. Mammogram revealed multicentricity. Hence, MRM was done

Please note: In our country, modified radical mastectomy is more frequently done. However, an early carcinoma is being treated more and more by breast-conserving surgery nowadays. Radical mastectomy is obsolete now.

Management of axilla

- In early breast cancer with no clinically apparent nodes and if the disease is not multicentric, then sentinel lymph node (SLN) biopsy can be considered. Otherwise, axillary dissection is recommended. Following surgery adjuvant treatment should be instituted in all tumours > 1 cm. ***Presence of metastatic disease within axillary lymph nodes remains the best single marker for prognosis.***

Sentinel node biopsy

- Senital means guard. Senital node is the first lymph node to get enlarged in malignancies.
- What does it mean? If no node is detected in this procedure in the axilla, axillary block dissection is not required. This will be of great significance in early breast carcinomas wherein lymph nodes are not clinically palpable nor detected by investigations such as ultrasound/CT scan of the axilla.
- Indication: Early breast cancer with (T1 or T2 No) clinically node negative axilla.

Contraindications

1. Palpable lymph node
2. Prior axillary sampling
3. Post-chemotherapy, post radiotherapy
4. Multifocal breast carcinoma
5. Allergy to blue dye

Localisation (procedure)

Peroperative is better and followed more commonly. Patent blue (Isosulfan vital blue dye 2.5–7.5 ml) or ^{99m}Tc radioisotope labelled albumin is used.

Where to inject

- Near the tumour: Peritumour area
- Into subdermal plexus around nipple.

Detection

- By visually blue staining after making an incision
- Hand held gamma camera. The node is biopsied and sent for frozen section biopsy or imprint cytology.

Where exactly to detect the sentinel lymph node (SLN)?

- A 2–4 cm incision is made in between the pectoralis major and lattissimus dorsi, after 5–7 minutes after the injection.
- Blue stained lymphatics are identified.
- 2–3 lymph nodes have to be removed.

Interpretation

- If sentinel lymph node (SLN) is negative for malignant cells, no axillary block dissection is required.
- If SLN is positive for malignant cells, axillary block dissection is done.

Detection rates

- Blue dye: 90%
- Radioisotope: > 95%.

Complications of MRM

1. *Seroma/lymph collection:* (30 to 50%). In spite of adequate drainage of the chest wall and axilla, drainage occurs for about 5–10 days. Just patiently reassure the patient.
2. *Secondary infection:* It manifests as redness, discharge, fever, etc. Appropriate antibiotics are necessary.
3. *Flap necrosis:* True mastectomy requires elevation of both upper and lower flaps (almost skin thin). Thus it predisposes to flap necrosis. This requires debridement, antibiotics, suturing and rarely skin grafting also.
4. *Haemorrhage:* Not common. If infection is severe especially in the axillary tissue, it is possible that the blood vessels may get eroded and bleeding can occur. This usually stops with pressure but may require ligation of bleeders in the operating theatre.
5. *Pain and numbness in the axilla, medial side of arm.* It is usually due to irritation of intercostobrachial nerve. Generally subsides by a few days of time. They require simple analgesics.
6. *Shoulder dysfunction* can occur especially when pectoral muscles are injured or retracted resulting in haematoma or when pectoralis muscles are removed. It improves over a period of time. Incidence is about 8–10%.
7. *Injury/thrombosis of axillary veins.* It manifests as severe pain in the hand and swelling. Treated by low molecular weight heparin.
8. *Injury to axillary vein*, needs to be repaired by 5 or 6–0 prolene sutures.
9. *Winging scapula* is due to injury to long thoracic nerve of Bell. The good anatomical knowledge is essential to prevent this complication.
10. *Lymphoedema* of the arm appears a few months later.

Adjuvant treatment—2 types

A. Radiotherapy (Key Box 21.19)

- Following either MRM or BCS, the residual disease is controlled by administering radiotherapy in a dose of 50 gray in 25 fractions.
- In the absence of nodal involvement axilla may be spared radiation.
- If 4 or more nodes are involved, then the field of radiation should include supraclavicular and internal mammary chains.
- Boosts to lumpectomy causes are given by adding 10–16 grays

KEY BOX 21.19

INDICATIONS FOR POSTOPERATIVE RADIOTHERAPY

- Tumour margin is positive
- Pectoralis major is involved
- Inner quadrant tumour
- High grade tumours
- Axillary clearance not satisfactory
- Breast conservative surgery
- Tumour size more than 5 cm, 4 or more nodes are positive

Brachytherapy in breast cancer (Figs 21.80 and 21.81)

1. It refers to use of radiation sources in or close to the tumour.
2. Breast is one of the organs which can be treated with brachytherapy.

- Details about brachytherapy are given in Chapter 47.

B. Systemic therapy—2 types

I. Adjuvant hormonal therapy

Hormonal therapy is administered to patient in whom the tumour expressed steroid hormone receptors, i.e. ER/PR.Those who are negative for ER/PR, will not benefit from these drugs.

The drugs are categorised as follows

- **1st line—anti-oestrogens**—tamoxifen.**Commonly, tamoxifen in a dose of 20 mg is used for 5 years, to be started only after completion of chemotherapy**. It is given in premenopausal patients.
- **Raloxifene** is an oral selective oestrogen receptor modulator **(SERM)** that has oestrogenic actions on bone and anti-oestrogen actions on the uterus and breast. It is used in the prevention of osteoporosis in postmenopausal women. Studies have proved that no specific advantage of raloxifene in the adjuvant treatment of breast cancer rather than established drugs such as tamoxifen.
- **2nd line—Aromatase inhibitors**. They prevent synthesi of endogenous oestrogens/steroids *by blocking th aromatase enzyme which converts androstenedione t oestradiol in the adrenals. They are: 1st generation*— aminoglutethimide and *2nd generation*—anastrozole letrozole, exemestane given to postmenopausal patients.
- **Letrozole** is costlier than tamoxifen, reduces oestroge levels by 98%, and thus slows down oestrogen sensitiv breast cancers. It is given in a dose of 2.5 mg per day. Side effects include vaginal bleeding, vaginal dryness, nigh sweats, hot flushes, osteoporosis, etc. (Key Box 21.20).
- **3rd line**—progestogens—megestrol acetate 400 mg per da can be given
- **4th line**—androgens such as fluoxymesterone in the dos of 30 mg daily is another drug (last 2 drugs are rarely given)

II. Adjuvant chemotherapy

Should be considered in all cases of early breast cance irrespective of menopausal status, hormone receptor statu and nodal status.

KEY BOX 21.20

AROMATASE INHIBITORS

- Aromatase is an enzyme which synthesises oestrogen
- It converts androgens into oestrogen by process of aromatisation
- Aromatase inhibitors inhibit this process
- Thus in premenopausal women—it can be used to decrease production of oestrogen by ovary. However, tamoxifen is better
- In postmenopausal woman—oestrogen is produced in peripheral tissues such as fat, liver and muscle
- Hence, drug of choice in postmenopausal patients is an aromatase inhibitor
- Anastrazole, letrozole, exemextane are the drugs
- Cardiac problems, osteoporosis are side-effects.

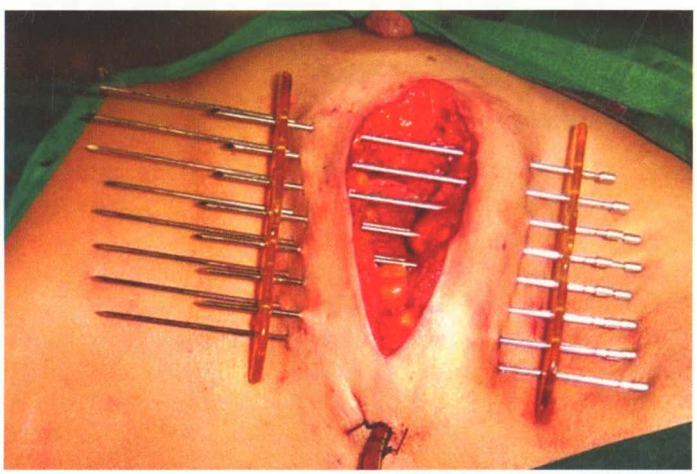


Fig. 21.80: Brachytherapy

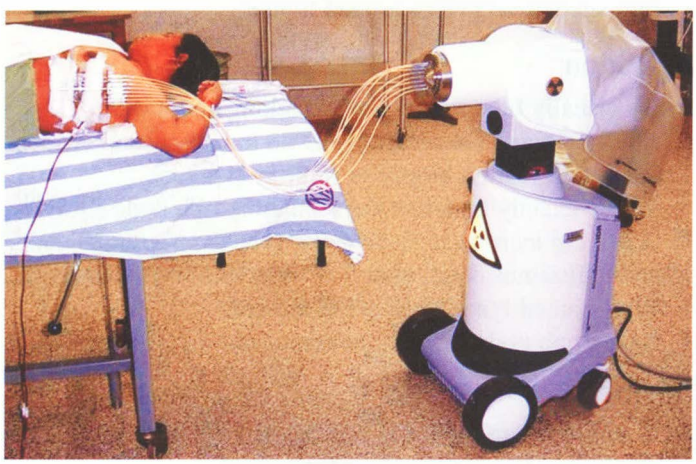


Fig. 21.81: Brachytherapy is being given

Classification of drugs

- **1st line agents:** Cyclophosphamide, adriamycin, 5-fluorouracil (FAC). Because of cardiotoxicity of adriamycin, **epirubicin** is preferred as in FEC regimen.
  The preferred regime is anthracyclines (adriamycin) which have a better response rate. Either CAF or FEC every 21 days × 6 cycles or at every 21 days × 4 cycles or just adriamycin with cyclophosphamide (AC). *However, the CMF (cyclophosphamide, mitomycin, fluorouracil) regimen still continues to be used widely due to economical reasons.*
- **2nd line agents:** Taxanes—paclitaxel and docetaxel
- **3rd line**: Gemcitabine

Indications for adjuvant chemotherapy

- Tumour > 1 cm
  OR
- Tumour < 1 cm with ER –ve, HER-2 +ve, high grade.

Adjuvant trastuzumab therapy

Trastuzumab: A monoclonal antibody against tyrosine kinase receptor (HER-2 receptor) is administered in patients with HER-2 +ve patients since it has been shown to improve disease free survival (DFS) by 50%, when it is combined with taxane-based chemotherapy. It potentiates effects of chemotherapy.

Dose: Loading 4 mg/kg.

Maintenance 2 mg/kg/week for 9 weeks.

Treatment of early carcinoma of the breast is summarised in Fig. 21.82.

LOCALLY ADVANCED BREAST CANCER (LABC)

Many patients with breast carcinoma come to the hospital with gross skin involvement, pectoral muscles involvement or chest wall involvement. Even though the cure rates are low, with proper planning, good control of the disease, good palliation in majority of patients and 'cure' in a small group can be achieved. They do not have metastasis. Hence, grouped under locally advanced breast carcinoma (T3 and T4). **With neoadjuvant chemotherapy (NACT), survival rates have improved** (Figs 21.83 and 21.84).

Aim

- Good locoregional control
- Attempt at 'cure' by chemotherapy.

Salient features of LABC (Figs 21.83 to 21.87)

- Any tumour more than 5 cm with or without skin and chest wall involvement are included under LABC.
- Under TNM, any stage IIB and stage III can be included under LABC. Isolated supraclavicular metastasis is also included in the stage III/LABC category.
- Mastitis carcinomatosa (without metastasis) is also included.

Types

A. Operable LOBC (large operable breast cancer) (IIB and III A): Ulceration, limited skin oedema, fixation to the pectoralis muscles and bulky axillary nodes are grave signs but **resection can be done.**

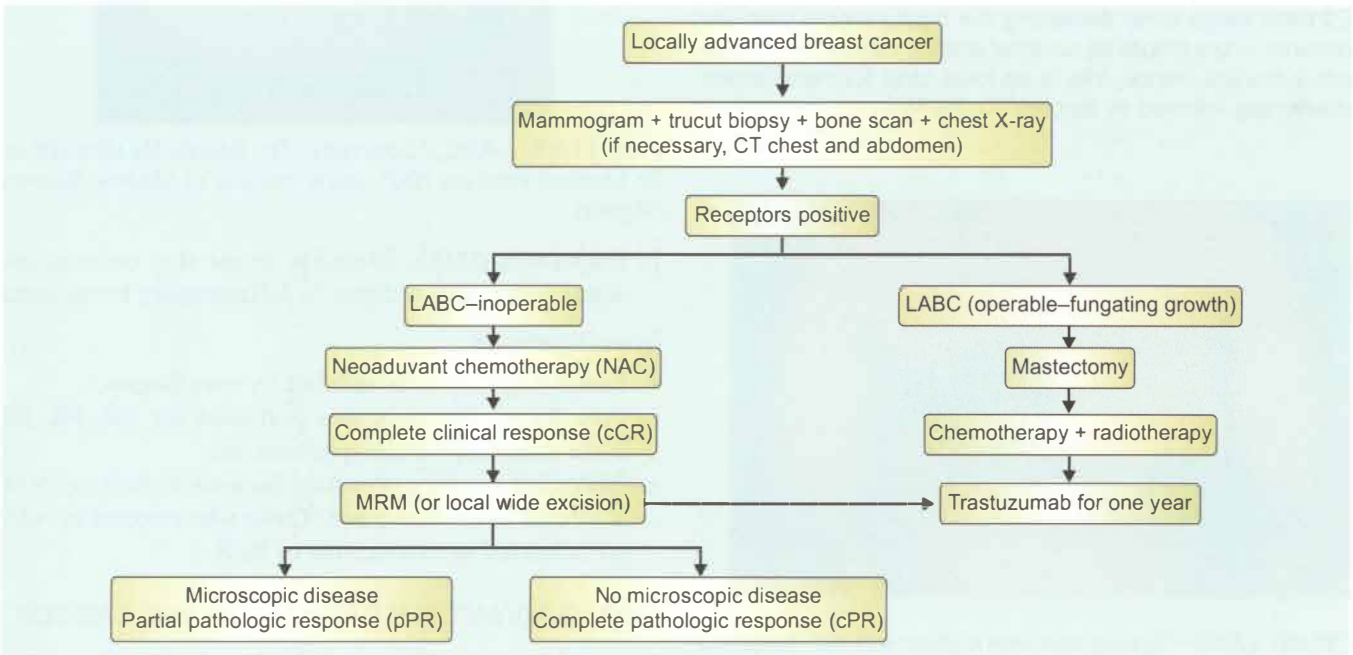


Fig. 21.82: Treatment of early carcinoma of the breast

Fig. 21.83: Carcinoma of the breast—See the puckering of the skin, nipple retraction—Core biopsy or incision biopsy also can be taken here. This case is not ideal for resection. Hence, neoadjuvant chemotherapy should be given first (T4 N1 M0)

Fig. 21.84: Large lump displacing the nipple areola complex downwards—core biopsy for receptor studies. MRM may not give negative margin. Hence, this is an ideal case for neoadjuvant chemotherapy followed by surgery (T4 N1 M0)

Fig. 21.85: LABC—Tumour was fixed to chest wall with fungating ulcer and oedema arm. (*Courtesy:* Dr Ankur Sharma, Assistant Professor, Dept. of Surgery, KMC, Manipal)

Fig. 21.86: LABC—tumour was fixed to skin with *peau d'orange* tumour was 8 cm in size (*Courtesy:* Dr Basavaraj Patil, Associate Professor, Dept of Surgery, KMC, Manipal)

Fig. 21.87: LABC (*Courtesy:* Dr Satish Deshmukh and Dr Murtaza Akhthar, NKP Salve Institute of Medical Sciences, Nagpur)

B. Inoperable (IIIB): Extensive breast skin oedema, intercostal nodes, arm oedema, or inflammatory breast cancer.

Investigations

- Tissue diagnosis is established by **core biopsy**.
- Core biopsy tissue is also evaluated for ER, PR, HER 2/neu status, P53 overexpression, etc.
- Biological markers important because if positive, NACT is offered in the first place. Those who respond to NACT, are subjected to mastectomy or BCS.

NEOADJUVANT THERAPY IN BREAST CANCER

This modality is used for treated locally advanced breast cancer (LABC) wherein due to the advanced nature of the disease in

the breast and/or axilla, the surgeon is unable to operate upon the patient initially. The exact TNM stage for defining LABC remains controversial (II B versus III A). However, any breast cancer in which the operating surgeon feels that he will be unable to completely resect the tumour in the breast and/or axilla can be treated with neoadjuvant chemotherapies.

If the growth becomes impalpable clinically after finishing ALL rounds of chemotherapy, it is called a **Complete Clinical Response** (cCR). However, if the resected specimen shows viable microscopic disease in a patient with cCR, it is termed **Partial Pathological Response** (pPR). If no microscopic growth is seen in resected specimen, then it is termed **Complete Pathological Response** (cPR).

The various modalities available are (Key Box 21.21)

1. **Neoadjuvant chemotherapy**
   - Patient should have a **good general condition** to receive chemotherapy.
   - Full course of chemotherapy should be completed whether or not the growth has completely resolved, e.g. 4 cycles of AC followed by 4 cycles of paclitaxel followed by surgery.
   - Patients who have received full course of neo-adjuvant chemotherapy before surgery need not receive any more chemotherapy in the adjuvant setting.
   - However, patients who did not receive full course of neoadjuvant chemotherapy before surgery should finish their remaining cycles after surgery.
   - Anti-HER2 (tratuzumab) can be given alongwith in the neoadjuvant setting till all courses of chemotherapies are over and should be continued in the adjuvant setting for a total duration of one year.
2. **Neoadjuvant hormonal therapy**
   - Postmenopausal patients who are not fit to receive systemic cytotoxic therapies should undergo a trial of this modality.
   - A prior assessment of the ER/PR status should be done to make sure that the tumour is hormone responsive. This modality does not work in hormone negative tumours.
   - Tamoxifen/letrozole/anastrozole all can be used.
   - Duration of the treatment should be up to the achievement of maximal response.
3. **Neoadjuvant radiotherapy**
   - Patients not responding to the above mentioned treatment may be given a trial of this modality.
   - Neoadjuvant therapy apart from making inoperable tumours operable also act as ***prognostic marker*** as in patients who achieve cPR have a far better outcome of the disease that those who do not.
   - Even in LABC, if the response to treatment is favourable **breast conservation is possible,** even in T4 tumours.

KEY BOX 21.21

SUMMARY OF LABC

- Trucut or open biopsy is done to confirm the diagnosis followed by **identification of receptor** status.
- Neoadjuvant chemotherapy is given first which will **shrink the tumour**.
- **Surgical resection** is carried out to achieve a clear margin (mastectomy or breast conservation surgery).
- **Radiotherapy** is given to chest wall and supraclavicular area.
- **Additional chemotherapy** can also be given (Adjuvant).
- If the tumour is fungating and the surgeon feels adequate cancer clear margin can be achieved, toilet mastectomy can be done first followed by chemoradiotherapy.

PEARLS OF WISDOM

- ***ER/PR negative responds better with NACT.***
- ***ER/PR positive has the option of NAHT (Neoadjuvant hormonal therapy).***
- ***HER-2/neu positive responds to Trastuzumab with taxanes, and response is better (treatment is costly)***

Advantages of neoadjuvant chemotherapy

- Downstages the disease
- Increases chances of breast conservation.
- Inoperable tumours may become operable.
- Systemic treatment (chemotherpy) starts early
- Assess response *in vivo*.
- Inhibits a potential postsurgical growth spurt.
- Chemotherapy is delivered through an intact vasculature.

CT Regimens

- Anthracycline (doxorubicin, epirubicin) + cyclophosphamide (AC/EC) with or without addition of 5 fluorouracil is found to be **superior to CMF regimen.**
- Addition of taxanes has been beneficial.
- **One example of sequential AC + Taxane (HER-2/neu negative case is given below:**
  Doxorubicin 60 mg/m^2—IV, and Cyclophosphamide 600 mg/m^2 IV day 1, cycled every 21 days for 4 cycles followed by Paclitaxel 175 mg/m^2 by 3 hour IV injection day/cycled every 21 days for 4 cycles.

Radiation therapy after LABC

RT is given to the chest wall in case of:

1. Tumour size > 5 cm before neoadjuvant therapy
2. Positive margins after mastectomy/BCT
3. More than 4 axillary LN +ve if axillary dissection done
4. Lymphovascular invasion
5. ER—negative
6. High grade tumour.

PEARLS OF WISDOM

Any recurrence following lumpectomy—mastectomy has to be done.

Role of "toilet" total mastectomy

- Controversial: It is not mentioned in most current literature.
- Fungating and offensive smelling—for local control to improve quailty of life and alleviate social and psychological factors in patients with poor physiological reserve.
- 30– 40% local recurrence.
- Increased incidence of distant metastasis without systemic therapy.
- Increased rates of wound dehiscience, wound infection, seromas—when associated with preoperative radiotherapy

Treatment of metastatic carcinoma of the breast

- Breast cancer is a systemic disease at the onset. Even at early stages, patient may harbour micrometastasis at some sites, which may remain dormant for years together or may get treated with adjuvant therapy. Metastatic breast cancer (MBC) may be seen in almost all organs but some organ involvement is seen more frequently than others. Soft tissue recurrence, bone, liver and lung metastasis is seen more commonly. Metastasis to brain, pleura and pericardium, altogether less common, may have devastating consequences and require specific treatment.
- Aim of the treatment is to give palliation in the form of treatment of symptoms.
- As these patients have advanced disease/disseminated disease, chemotherapy is given first followed by excision of lump/simple mastectomy to get rid of fungating tumour. However, if the **lump disappears totally, there is no indication for excision** of the lump or mastectomy.
- Following chemotherapy, tamoxifen is given for a period of five years. Repeated courses of second and third line chemotherapeutic drugs may have to be given.
- The treatment of individual organ involvement or system involvement is given below.

Patients with oestrogen receptor +ve status are more likely to have a bony recurrence, whereas ER –ve tumours tend to recur in liver and brain. After diagnosis of MBC, patients live an average of 16–30 months.

Treatment options in MBC

1. Hormonal treatment
2. Chemotherapy
3. Trastuzumab
4. Supportive treatment.

Hormonal treatment

In general, hormone treatment is better tolerated as it is associated with fewer side effects. Hence, it is preferred in patients who have steroid receptor positive status and/or asymptomatic visceral metastasis.

Chemotherapy

- Preferred in patients with steroid receptor negative disease and in symptomatic visceral metastasis.
- Both single and combination chemotherapy regimen can be used.

Trastuzumab

- Patients who have overexpression of HER-2 receptor, may also receive trastuzumab in addition to the above.

Supportive treatment—3 organs

1. Malignant pleural effusion

- Once effusion is confirmed by aspiration and cytology, an intercostal drain is left in place for a few days. Once the drainage is nil or completely dry, talc insufflation is done, to achieve **pleurodesis**. Talc is the most effective agent, followed by tetracycline or bleomycin, etc. In selected cases, effective pleurodesis has given an asymptomatic period up to 1–2 years.

2. Cerebral metastasis

- Patients present with features of raised intracranial pressure such as headache, nausea, vomiting and papilloedema.
- Treatment includes **corticosteroids** and cranial radiotherapy. However, treatment is distressing.

3. Bone metastasis

- Eventually 60–70% of patients with carcinoma breast develop bone metastasis. Bone metastasis produces intractable pain, pathological fractures, quadriplegia and paraplegia, etc. (Figs 21.88 and 21.89 and Key Box 21.22).
- Localised bone lesions are treated by palliative radiotherapy or decompression as in quadriplegia, etc.
- **Bisphosphonates**, e.g. oral clodronates have been found to arrest progression of bone disease, given at the dose of 1,600 mg/day (Key Box 21.23).

Causes of death

- Malignant pleural effusion
- Bony metastasis
- Cerebral metastasis
- Cancer cachexia.

Management of metastatic breast cancer is summarised in Fig. 21.90.

KEY BOX 21.22

BONE METASTASIS

- Pain—due to stretching of periosteum
- Pathological fractures
- Paralysis
- Perish—due to hypercalcaemic crisis

Fig. 21.88: Pathological fracture humerus—treated by internal fixation followed by radiotherapy

KEY BOX 21.23

BISPHOSPHONATES

- Important drugs in addition to radiotherapy to treat bone metastasis in malignancies.
- Used to treat hypercalcaemia.
- Inhibits osteoclast formation
- Decreases bony resorption and induces apoptosis of osteoclasts
- Decreases bony pain, increases mobility
- Helps in recalcification of lytic bone metastasis
- IV—pamidronate or zoledronic acid 4 mg IV monthly for 10 courses are used

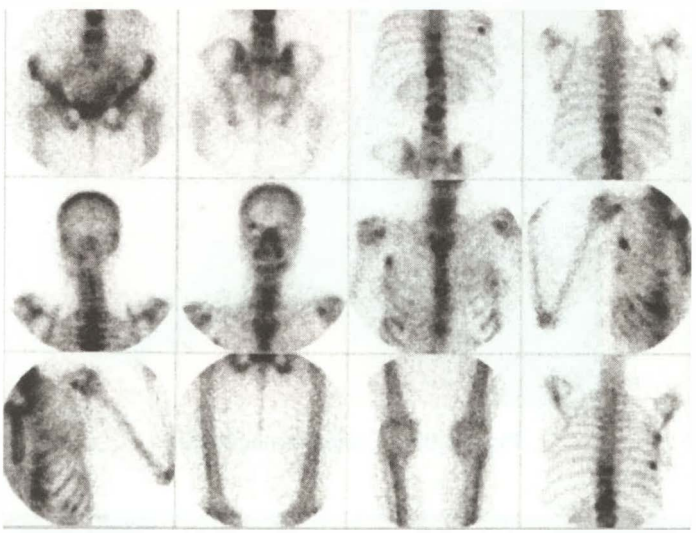


Fig. 21.89: Whole body bone scan showing disseminated bony metastasis—^{99m}Tc-labelled phosphate compounds are agents of choice for detection of osseous metastasis

See Fig. 21.90 for flow chart of management of metastatic breast cancer

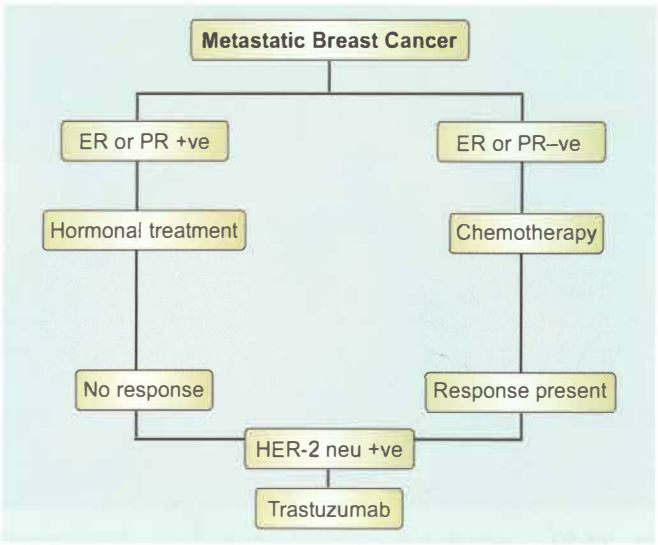


Fig. 21.90: Management of metastatic breast cancer

EFFECT OF LYMPHATIC OBSTRUCTION FROM CARCINOMA OF BREAST

1. ***Peau d'orange:*** It occurs due to lymphoedema due to subareolar lymphatic obstruction and fixation of hair follicle to the underlying malignancy.
2. Oedema of the arm is also called ***elephantiasis chirurgens*** (Figs 21.91 and 21.92).
   - It is a complication of radical mastectomy when all the nodes in the axilla are removed, especially when postoperative radiotherapy is given in the axilla.
   - It occurs due to destruction of all lymphatics and lymph nodes.
   - It does not pit on pressure. Some amount of infection also plays a role.

   Treatment
   - Difficult. Elastic bandage, exercise, massage and antibiotics may help.
3. **Brawny oedema of the arm**
   - This occurs in inoperable carcinoma of breast with fixed nodes in the axilla.
   - Arm is oedematous and does not pit on pressure.
   - It is brawny, indurated, painful.
   - It is treated like above.
4. **Cancer-en-cuirasse** (Figs 21.93 to 21.95)
   - In this, the entire chest wall is studded with cancerous nodules which are hard and fixed to the skin and to the chest wall. The condition has been compared to an armour used by the soldier. It indicates advanced carcinoma of breast. Palliative treatment is given in the form of radiotherapy or chemotherapy and analgesics to relieve the pain.
5. **Lymphangiosarcoma:** It is a rare complication after mastectomy and arises from lymphoedematous limb. It is treated by forequarter amputation.

Fig. 21.91: Lymphoedema right upper limb—troublesome complication after axillary block dissection—more after radiotherapy to axilla

Fig. 21.92: Postmastectomy lymphoedema (*Courtesy:* Dr Sreejayan, Professor of Surgery, Calicut Medical College, Calicut)

Fig. 21.93: Cancer-en-cuirasse—Chest wall studded with cancerous nodules—very painful bleeding lesion

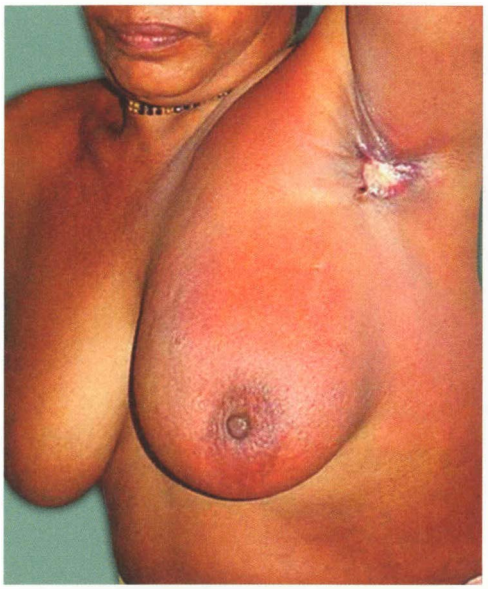


Fig. 21.94: Extensive lymphoedema and lymphangitis due to carcinoma axillary tail of the breast

Fig. 21.95: Cancer-en-cuirasse

BREAST RECONSTRUCTION

- The ideal candidate for breast reconstruction is a patient who has undergone modified radical mastectomy.

Mastectomy results in following changes in a woman such as:

- Psychological stress
- Mood disturbances and anxiety

- Increased consciousness about clothes
- Decreased sexual interest and satisfaction
- Negative body image

Reconstruction (Key Box 21.24)
- Improves self confidence
- Better social life
- Decreases concern about cancer
- Better sexual life
- Feel "whole again"

Timing: Immediate or delayed
- Immediate reconstruction: Should be done if no contraindications. It has proven psychological benefits and patient satisfaction, it is cost effective, it does not delay adjuvant treatment and recurrence detection.

KEY BOX 21.24

COMPONENTS OF BREAST RECONSTRUCTION

- **Chest wall reconstruction:** This can be achieved by using latissimus dorsi musculocutaneous flap or contralateral transversus abdominis muscle (TRAM) flap.
- **Creation of a mould:** A silicone gel implant is inserted under pectoralis major muscle (subpectoral pocket).
- **Reconstruction of nipple and areola**: This can be achieved by skin taken from inner thigh, labia minora or from opposite breast (nipple sharing). However, this reconstruction is done 4–6 weeks later, once the implant settles down.
- **Symmetry with the opposite breast**: It is achieved by doing reduction mammoplasty of the other breast.

Reconstruction options

1. **Pedicled flaps**
   - Latissimus dorsi myocutaneous flap (with implant)
   - TRAM flap
2. **Free flaps**
   - TRAM flap
   - Gluteus maximus myocutaneous flap
   - Anterolateral thigh flaps
3. **Silicon compound gel/saline**
   - Expandable
   - Adjustable
   - Lesser risk of complications

BREAST RECONSTRUCTION WITH TRAM FLAP (Figs 21.96 to 21.101)

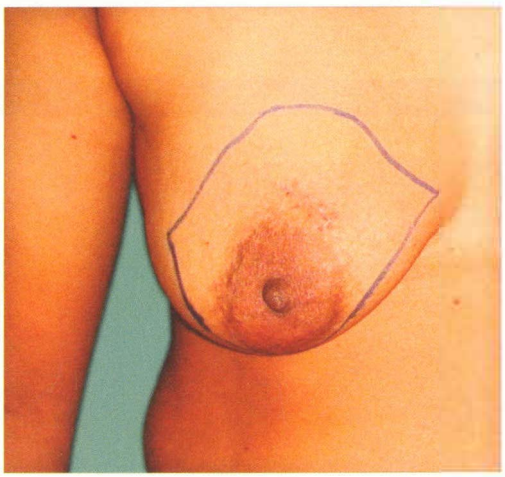


Fig. 21.96: Mastectomy site is marked

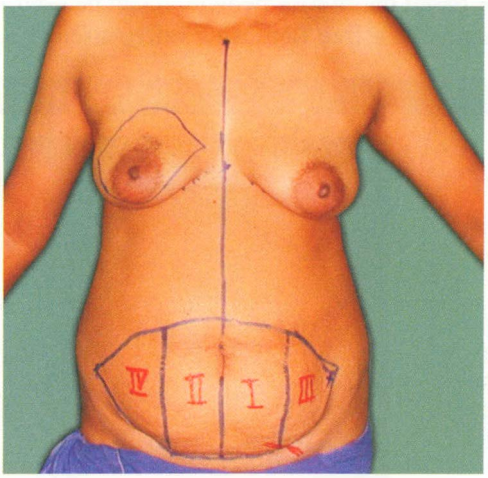


Fig. 21.97: TRAM flap marking is done

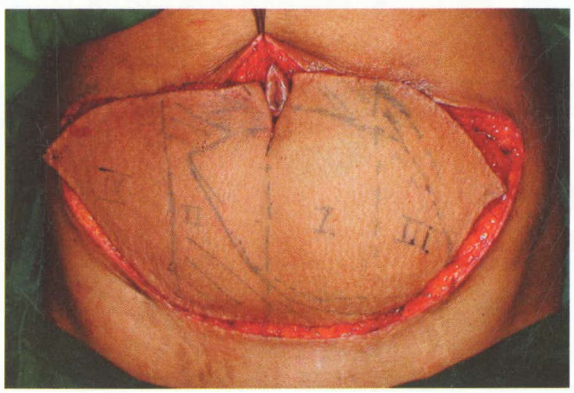


Fig. 21.98: TRAM flap is raised—flap design: Zone I–IV

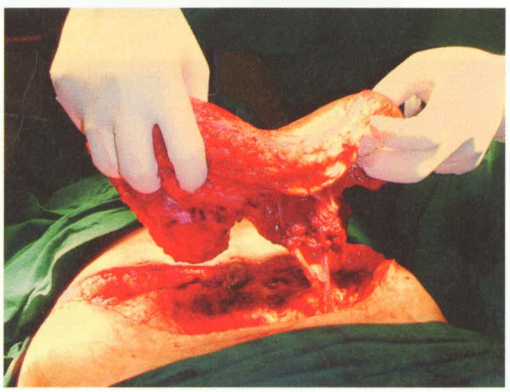


Fig. 21.99: TRAM flap is shown with its blood supply

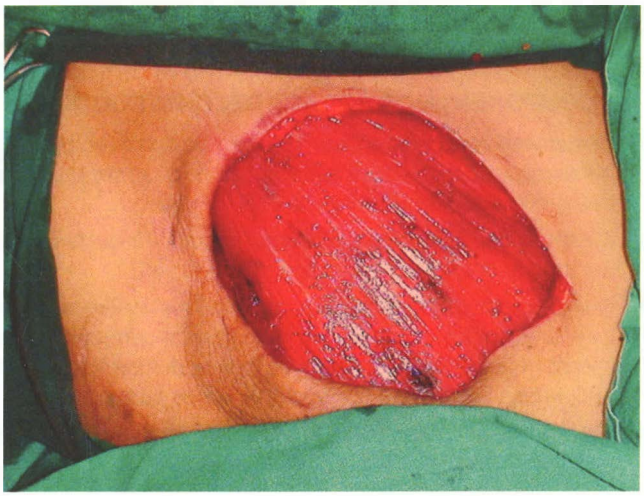


Fig. 21.100: Mastectomy bed—pectoralis muscle fibres

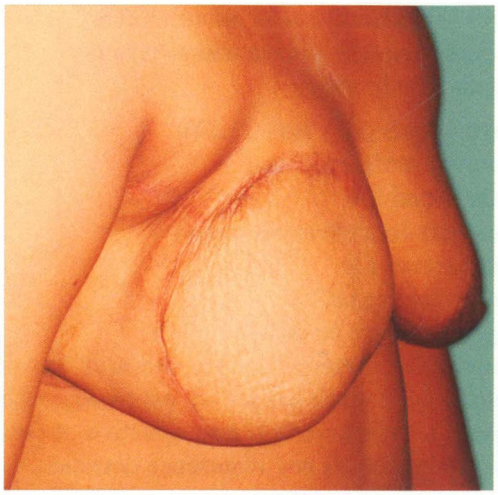


Fig. 21.101: Healing after 20 days

(*Courtesy:* Dr Bhaskar KG, Senior Consultant, Plastic Surgeon, Medical Trust Hospital, Cochin, Kerala (next page photos also)

Breast reconstruction with TRAM flap

Transversely oriented rectus abdominis myocutaneous flap popularly known as **TRAM flap is the most common reconstructive option in postmastectomy breast reconstruction.** This flap can be pedicled based on the superior pedicle or based on inferior pedicle. It can be used for breast reconstruction as a free flap using microvascular technique.

Superior epigastric artery is the artery supplying the flap. If it is used as **a pedicled flap** for breast reconstruction, the contralateral side is used for reconstruction.

If TRAM flap is taken with inferior pedicle using microvascular technique, deep inferior epigastric artery forms the main vascular basis of the flap.

BREAST RECONSTRUCTION WITH LD FLAP (Figs 21.102 to 21.104)

Fig. 21.102: Latissimus dorsi (LD) flap is marked

Fig. 21.103: LD flap is raised

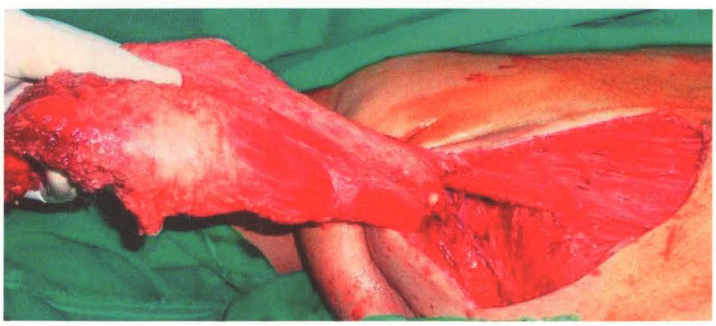


Fig. 21.104: LD flap is brought to the mastectomy site

BREAST RECONSTRUCTION WITH LD FLAP WITH SILICON IMPLANT (Figs 21.105 to 21.108)

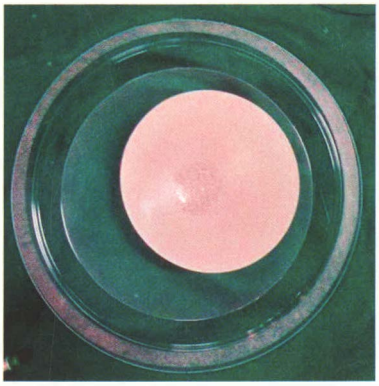


Fig. 21.105: Silicone implant

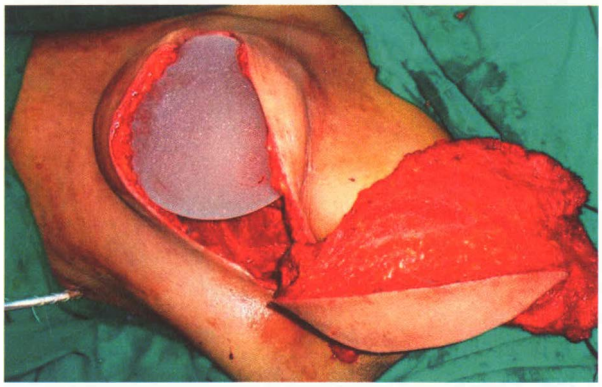


Fig. 21.106: Silicone implant placed

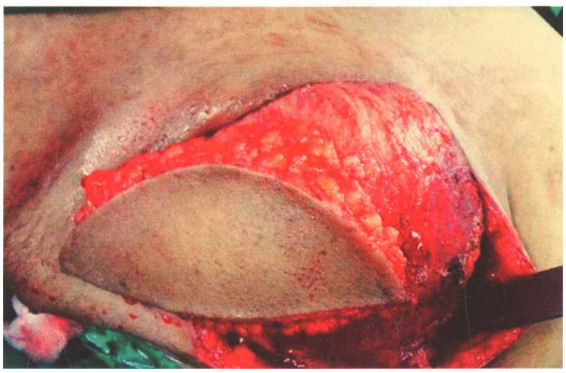


Fig. 21.107: Silicone implant placed in the subpectoral pocket

Fig. 21.108: Wound closed after implant

Latissimus dorsi musculocutaneous flap

Indications: Breast or skin reconstruction, post mastectomy or wide excision of large chest wall

Contraindications:

1. Previous lateral thoracotomy
2. Very large breast in patient who does not desire reduction
3. Planned postoperative radiation therapy

Positioning:

- Lateral decubitus position
- Marked before mastectomy

Vasculature/Blood supply: Thoracodorsal pedicle

Procedure: Skin island of about 5–6 cm wide, outlined transversely. Divide the paraspinal origins of the muscle preserving the pedicle. Rotated on its humeral insertion towards the anterior chest wall beneath a bridge of thoracic skin below the axilla. Breast implant can be placed in a submuscular pocket.

Advantages:

1. Proximity to the breast
2. Reliable/robust circulation (rich blood supply)

Disadvantages:

1. Large scar on the back.
2. Cannot be used if there is an injury to thoracodorsal pedicle.

MALE BREAST CARCINOMA

Introduction

Constitutes less than 1% of all cancer in men. Minimum age is around 65 years

Probable aetiological factors

1. Radiation exposure
2. Oestrogen therapy
3. Gynaecomastia
4. Klinefelter's syndrome XXY
5. Testicular feminising syndrome
6. Obesity
7. Increases incidence in prostate cancer patients
8. Bantumen

Pathogenesis

Male breast cancers tend to be more advanced at the time of presentation because of less subcutaneous fat. More than 90% cases are infiltrating duct carcinoma, about 10% are ductal carcinoma *in situ* (DCIS).

Fig. 21.109: Male breast carcinoma in pre-existing gynaecomastia. You can see destruction of the nipple and infiltration into the skin: Commonly it is infiltrating ductal carcinoma

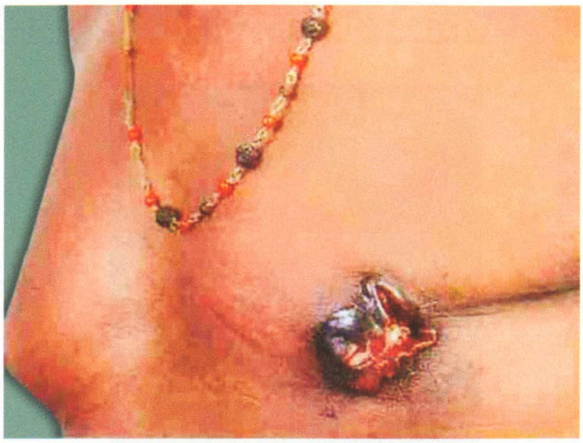


Fig. 21.110: Male breast carcinoma—tends to ulcerate and infiltrate chest wall early (*Courtesy:* Dr Surya Prakash Saxena, District Surgeon, Durg, Chhattisgarh)

Clinical features (Figs 21.109 and 21.110)

- Lump in the breast which is hard and irregular.
- Nipple retraction
- Early skin involvement
- Metastatic axillary nodes.

Differential Diagnosis

- Chest wall sarcoma
- Bony lesions other bony metastasis, tuberculosis
- Gynaecomastia.

Investigations

- FNAC/Trucut biopsy
- Chest X-ray.

Treatment

- MRM with radiation to the chest wall
- 80% of these cases are ER positive, hence role for tamoxifen
- Goserelin 3.6 mg/28 days for 2 years—it is a LHRH agonist. It causes reversible chemical castration.

MISCELLANOUS

PROPHYLACTIC MASTECTOMY

It is also called 'risk reduction mastectomy'.

Counselling before risk reduction mastectomy

- Loss of breast is irreversible
- Sexual and psychological function
- Reconstruction has its complications
- Does not guarantee 100% safety against cancer.

Cancer risks in *BRCA* mutation carriers

- *BRCA* 1: 65 to 85% (long arm of chromosome 17)
- *BRCA* 2: 40–85% (long arm of chromosome 13)
- *BRCA* mutations: Highest incidence in Ashkenazi Jews
- High risk of ovarian cancer
- High risk of contralateral breast cancer
- Multiple ipsilateral primaries
- Adenocarcinoma of invasive type
- High grade
- *BRCA* 1—ER, PR, HER-2-neu negative
- High risk for a recurrence.

Aim

- To decrease the risk of developing invasive cancer
- Also to decrease the risk of dying.

Risk assessment

2 specific genes—*BRCA* 1 and *BRCA* 2 are responsible for 5% of all breast cancers.

Why prophylactic mastectomy

- *BRCA* 1 associated cancers are invasive, high grade, ER, PR and HER-2/neu negative. Hence, more aggressive and carry poor prognosis.

Other risk reduction strategies

- Lifestyle modification: Exercises, decrease alcohol intake, avoid HRT.
- Early detection by annual mammogram
- Tamoxifen as a primary chemoprevention drug.

Types of risk reduction mastectomy

- Total mastectomy
- Subcutaneous mastectomy.

Indications (Pennisi, 1986)

1. Biopsy proved
   - Noninvasive intraductal carcinoma *in situ*
   - A single focus of lobular carcinoma *in situ*
   - Florid cystic disease
2. A mammogram that is suspicious of moderate to severe mammary dysplasia

3. Persistant breast nodules that do not vary with menstrual cycles and are of concern to the patient and physician
4. Familial or hereditary breast carcinoma.

Procedure

- Submammary incision is given.
- Breast tissue is dissected from the pectoral muscles.
- Skin flap along with even thickness of subcutaneous fat which is retained under the skin, is raised (helps in retaining vascularity of the flap). Haemostasis is obtained. Immediate reconstruction or reconstruction at a later date can be done.
- Skin sparing mastectomy
- **Remove entire breast including axillary tail.** Flaps should be thin. Axillary block dissection is not necessary.

Results

- Prophyhlactic mastectomy reduces incidence of breast cancer by 90%.

MONDOR'S DISEASE

Mondor's disease is spontaneous thrombophlebitis of the superficial veins of the breast and anterior chest wall. The common causes of thrombophlebitis such as injury or infection are not found in these cases.

Signs

1. Indurated subcutaneous thrombophlebitic cord about 3 mm diameter of varying length is situated in the subcutaneous plane with an attachment to the skin. Consistency is like that of vas deferens.
2. When the skin over the breast is stretched by raising the arm, a narrow, shallow subcutaneous groove along the side of the cord becomes apparent.

Treatment

- Restricted arm movements (otherwise it is very painful). Spontaneous recovery is expected within a few days.

ANGIOSARCOMA OF THE BREAST

- They are uncommon malignant tumours of the breast.
- Aetiology is not clear
- Breast is one of the sites of angiosarcoma.
- Clinically it presents as rapidly growing lump in the breast with infiltration of the skin producing ulceration.
- On careful observation, the edges are not everted and the lesion is very vascular.
- It is difficult to make a clinical diagnosis of angiosarcoma of the breast (Fig. 21.111). However, with careful examination it is possible to suspect because of increased vascularity of the tumour and the features mentioned above.
- Confirmation of the diagnosis is by FNAC or by biopsy from the lump. Chest X-ray and CT scan of the chest are other helpful investigations.

Fig. 21.111: Angiosarcoma of the left breast

- Treated by course of chemotherapy followed later by mastectomy.

DISORDERS OF AUGMENTED BREAST

- This is called augmentation mammoplasty
- Implant used is outer silicone shell filled with silicon gel, saline or a combiantion of both
- Site of implant

A. **Subpectoral:** More often this is the side. Even if a cancer develops in this breast, mammography can detect early lesion. After mastectomy—if recurrence develops, easy to detect if it is subpectoral

B. **Subcutaneous:** Not many advantages: Easy to keep and easy to remove. It interferes with detection of malignancy. Easy to rupture.

- **Complications**
  1. Implant rupture: 5 to 10%
  2. Bleed

RARE BREAST CANCERS

1. **Squamous cell (epidermoid) carcinoma**
   - Rare cancer from metaplasia within duct system and devoid of distinctive clinical/radiological characteristics.
   - Regional metastasis occurs in 25% of patients
   - Distant metastasis is rare
2. **Adenoid cystic carcinoma**
   - Rare, less than 0.1%.
   - Well circumscribed and usually 1–3 mm in diameter.
   - Axillary lymph node metastasis are rare
   - Death usually occurs from pulmonary metastasis.
3. **Apocrine carcinoma**
   - Well circumscribed cancers have rounded vesicular nuclei and prominent nucleoli.
   - Usually aggressive.

4. **Sarcomas**
   - Large painless breast mass with rapid growth
   - Diagnosis: Core/open biopsy.
   - Sarcomas graded on—eccentricity, degree of differentiation, nuclear atypia, mitotic activity
   - Axilla is addressed only if palpable lymph nodes are present
   - Usually in patients with history of adjuvant RT.
5. **Lymphomas:** 2 variants (good prognosis)
   a. Women < 39 years—bilateral, associated with Burkitt's lymphoma. Axillary dissection is done for staging and clearance of the disease. Recurrence is treated with CT/RT
   b. Women > 40 years—B cell type. Rarely Hodgkin's lymphoma

AN EXAMPLE OF A CASE OF CARCINOMA BREAST

A 59-year-old post-menopausal lady complains of lump in the right breast of 3 months duration. Lump was hard in consistency. Sonomammogram revealed ill-defined hypoechoic lesion with spiculations seen in the superolateral quadrant of the breast. FNAC was suggestive of malignancy. Whole body bone scan after 3 hours IV inj. of 20 mCi of Tc-99 m MDP showed few osteoblastic lesions—so degenerative changes?

Treatment: She underwent local wide excision followed by axillary block dissection. Final report was tubular carcinoma—30% arising in a background of proliferative breast disease (70%). All margins were free. Lymph nodes were free from metastasis

Immunohistochemistry: Tumour cells are ER positive PR positive Quick score 2. Her-2/neu 1+ Ki 67: 12%—Hormone responsive

Next plan: External beam radiotherapy of 60 Gy in 30 fractions over 6 weeks, including boost to the lumpectomy side.

- She also received tablet letrozole 2.5 mg HS and 6 monthly Zoledronate — Zoledic acid

Based on DNA microarrays, breast cancer can be divided into specific groups referred to as luminals depending on which the choice of the treatment can be given. However, since these tests can be very expensive, IHC results (ER/PR/HER/Ki 67) can be used as surrogate markers to define various categories of breast cancer.

RECENT ADVANCES IN THE MANAGEMENT OF CARCINOMA BREAST (Table 21.9)

1. Luminal A

Tumour characteristics should include all of the following features:

ER and PR positive

HER-2 negative

Ki 67 'low' (i.e. <14%)

Recurrence risk 'low' based on multi-gene-expressior assay (Oncotype DX) i.e. < 25 score

2. Luminal B

'Luminal B-like (HER-2 negative)'

ER positive

HER-2 negative and at least one of:

Ki 67 'high'

PR 'negative or low' i.e. score less than or equal to 19%

Recurrence risk 'high' based on multi-gene-expression assay , i.e. > 25 score

'Luminal B-like (HER-2 positive)'

ER positive

HER-2 over-expressed or amplified

Any Ki 67

Any PR

3. Non-luminal (Erb B2 over-expression)

HER-2 over-expressed or amplified

ER and PR absent

4. Basal-like-triple negative (ductal)

ER and PR absent

HER-2 negative.

Duration and type of chemotherapy

1. Anthracycline based chemotherapies (e.g. doxorubicin) should be preferred over the first generation chemotherapy regimens like CMF.
2. Taxols should be added sequentially to anthracycline based agents especially when the tumour is HER-2 positive and/or more than 4 lymph nodes are positive or triple negative disease.
3. 6–8 cycles should be the number of cycles given to the patient.

   One example would be 4 cycles of adriamycin (doxorubicin) + cyclophosphamide given 3 weekly followed by 4 cycles of single agent paclitaxel given 3 weekly.
4. Duration of trastuzumab should be 1 year. It should not be given along with anthracycline based chemotherapy but can be given along with taxols.
5. Dose dense chemotherapies are the ones where the overall duration of chemotherapy cycles is reduced, e.g. instead of conventional three weekly gap between two cycles, the gap is reduced to 2 weeks. These are typically used in aggressive breast cancers like triple negative breast cancers.

Duration and types of hormonal therapies

1. Tamoxifen can be used in both pre- and post-menopausal ladies, the total treatment duration is **now recommended to be 10 years (earlier it was 5 years), especially when the chances of recurrence is high.**

Table 21.9 Other molecular subtypes may include varieties like 'molecular apocrine', 'normal breast like' , 'claudin low' .

Subtype	Type of therapy	Notes
Luminal A	Hormonal therapy (e.g. tamoxifen, letrozole, anastrozole)	Relative indications of adding chemotherapy to hormonal therapy include: 1. High recurrence score on oncotype DX, i.e. > 25 2. High score on MammaPrint 3. Grade 3 disease 4. Involvement of 4 or more axillary lymph nodes. 5. Age < 35 years
Luminal B (HER-2 negative)	Adjuvant chemotherapy followed by hormonal therapy	
Luminal B (HER-2 positive)	Adjuvant chemotherapy + anti HER-2 (trastuzumab) + hormonal therapy	Trastuzumab should be given in any tumour size greater than 5 millimeters
Nonluminal	Adjuvant chemotherapy + trastuzumab	Aggressive regimens like **DOSE DENSE** chemotherapy may be used.
Triple negative	Adjuvant chemotherapy	

2. Aromatase inhibitors are typically used in **post-menopausal ladies** and should not be used in pre-menopausal females unless it is given along with ovarian suppression agents like Goserelin.
3. Other agents **like Fulvestrant** should only be used in the second line setting.
   - SERMs: Selective estrogen receptor modulators (agonistic in some tissues while antagonist in others)
   - Example: Tamoxifen is agonist on bone and uterus but antagonist in breast. Other example: Raloxifen
   - Fulvestrant, on the other hand is a pure antagonist.

Novel agents in breast cancer

1. Trastuzumab—monoclonal antibody against HER-2/neu receptor on the breast cancer cell surface.
2. Lapatinib—dual tyrosine kinase inhibitor
3. Pertuzumab—HER dimerisation inhibitor
4. Ado-trastuzumab emtansine (T DM1)—antibody drug conjugate
5. Ixabepilone—microtubule stabilizer
6. Denosumab—a RANK ligand inhibitor used for palliation of bone metastasis.

INTERESTING 'MOST COMMON' IN BREAST DISEASES

- Most common organism in lactational mastitis is *Staphylococcus aureus.*
- Most common organisms in nonlactational mastitis are anaerobes.
- Most effective drug used to treat cyclical mastalgia is evening primrose oil.
- Most common cause of gynaecomastia is idiopathic.
- Most breast cancers arise in ductal epithelium (90%)
- Most breast cancers occur in upper outer quadrant (60%)
- Most common presentation of carcinoma breast is with a lump
- Most common type of carcinoma breast is scirrhous carcinoma (60 to 70%).
- Most malignant form of carcinoma breast is mastitis carcinomatosa.
- Most common reconstructive option in postmastectomy breast reconstruction is TRAM flap.
- Most commonly used hormonal treatment in carcinoma breast is tamoxifen

WHAT IS NEW IN THIS CHAPTER?/ RECENT ADVANCES

- All topics have been updated.
- The understanding of pathology and treatment of breast abscess has been updated.
- Prophylactic mastectomy, sentinal node biopsy, flap breast reconstruction, has been edited
- Latest treatment guidelines 'LUMINAL' has been added in carcinoma breast.

MULTIPLE CHOICE QUESTIONS

1. Following are true for lymphatic drainage of the breast *except*:
A. Apical nodes are also called infraclavicular nodes
B. Apical nodes drain into subclavian lymph trunk
C. Posterior third of the breast drain into supra-clavicular nodes
D. For detection of sentinel node, ideal site is subdermal plexus around nipple

2. Following are true for advantages of MRI in the breast *except:*
A. It is the best modality for women with breast implants
B. Screening in women with strong family history
C. It is also better than ultrasound to image axilla
D. It can distinguish scar from recurrence in women who have undergone breast conservative surgery

3. Following are true for retraction of the nipple *except*:
A. Slit-like retraction is seen in duct ectasia
B. Circumferential retraction is seen in carcinoma breast
C. Extension of the growth along lactiferous duct causes retraction of the nipple
D. Horizontal retraction can occur at puberty suggests fibroadenosis

4. Following are true Lactational mastitis *except*:
A. Retracted nipple is one of the cause
B. Majority of the cases are due to anaerobic infection
C. Repeated aspiration is recommended treatment
D. Fluctuation is a late sign

5. Smoking is associated with which of the following breast disease?
A. Tuberculosis B. Breast abscess
C. Duct ectasia D. Mondor's disease

6. Following is not the common sign of Periductal mastitis:
A. Discharge per nipple
B. Indurated mass
C. Fistula
D. Circumferential retraction of the nipple

7. The widely used first investigation of choice in a lady of 25 years with lump breast is:
A. Ultrasonography
B. CT scan
C. MRI
D. FNAC

8. Which one of the following is not the treatment for mastalgia?
A. Evening primrose oil
B. Danazol
C. Steroids
D. Bromocriptine

9. Following are true for Phylloides tumour *except*:
A. Usual age of presentation is 20 years
B. Large tumour with bosselated surface
C. It may have high mitotic index
D. Rarely develop into sarcoma

10. Which one of the following is the treatment of choice for early breast cancer in a 30-year-old lady who is 4 months pregnant?
A. Chemotherapy
B. Tamoxifen
C. Local wide excision
D. Modified radical mastectomy

11. Which is the drug used in a patient to prevent breast cancer with positive family history but unlikely carrier of breast cancer gene?
A. Adriamycin B. Tamoxifen
C. Letrozole D. Trastuzumab

12. Which one of this operative step is not done in Modified radical mastectomy?
A. Total mastectomy
B. Axillary block dissection
C. Removal of pectoralis major
D. Removal of pectoralis minor

13. Following are the components of breast conservative surgery *except*:
A. Excision of tumour plus a rim of 1 cm of normal breast tissue
B. Axillary block dissection
C. Routine removal of the skin over the tumour
D. Sentinel node biopsy in selected patients

14. Following are true for radiotherapy in carcinoma breast *except*:
A. It should be given after breast conservative surgery
B. Large tumours after surgery require radiotherapy
C. Extensive lymphovascular invasion is an indication
D. Routinely given to axilla after a complete block dissection

15. Following are true for aromatase inhibitors *except:*
A. Maximum use in the premenopausal patients
B. Relapse free survival is prolonged
C. Reduction in the contralateral disease
D. Increase in the bone density loss

16. While of the following is the most important aetiological factor for carcinoma breast?
A. Oral contraceptive pills
B. Childhood irradiation for Hodgkin's lymphoma
C. Duct ectasia
D. Racial factors

17. Following are true for Ductal carcinoma *in situ* (DCIS) *except:*
A. It is usually unilateral
B. Multifocal
C. Excision with or without radiotherapy is the treatment of choice
D. No role for chemotherapy

18. Following are true for inflammatory carcinoma *except*:
A. It results in blockage of dermal lymphatics
B. It is considered as locally advanced breast cancer
C. Neoadjuvant chemotherapy is the treatment of choice
D. Severe degree of inflammation present pathologically

19. Following are true for lobular carcinoma *in situ except*:
A. It is usually unilateral
B. Multifocal
C. Excision with or without radiotherapy is the treatment of choice
D. 20 % will develop into invasive carcinoma

20. Following are true about internal mammary lymph nodes *except*:
A. They drain posterior part of the breast
B. They communicate with subdiaphragmatic lymph nodes
C. They are included in the staging system now
D. Their involvement indicates metastatic disease

21. Following are true for sentinel node biopsy in carcinoma breast *except*:
A. Injection of patent blue localises the sentinel node
B. Injecting into subdermal plexus around the nipple is ideal
C. Hand held gamma camera detects this
D. It is ideal in clinically node positive axilla

22. Following is an indication for radiotherapy to chest wall in carcinoma breast *except*:
A. Extensive lymphovascular invasion
B. Large number of positive nodes in the axillla
C. Base is involved by the tumour
D. Skin is infiltrated

23. Following are true for tamoxifen *except*:
A. It is widely used as hormonal treatment in premenopausal patients
B. It decreases the annual recurrence by 25%
C. It also has benefit in preventing contralateral breast cancer
D. It is not recommended in patients who have family history of carcinoma breast but unlikely carriers of breast cancer gene

24. Treatment option for carcinoma breast in pregnancy is:
A. Local wide excision
B. Radiotherapy
C. Chemotherapy
D. Mastectomy

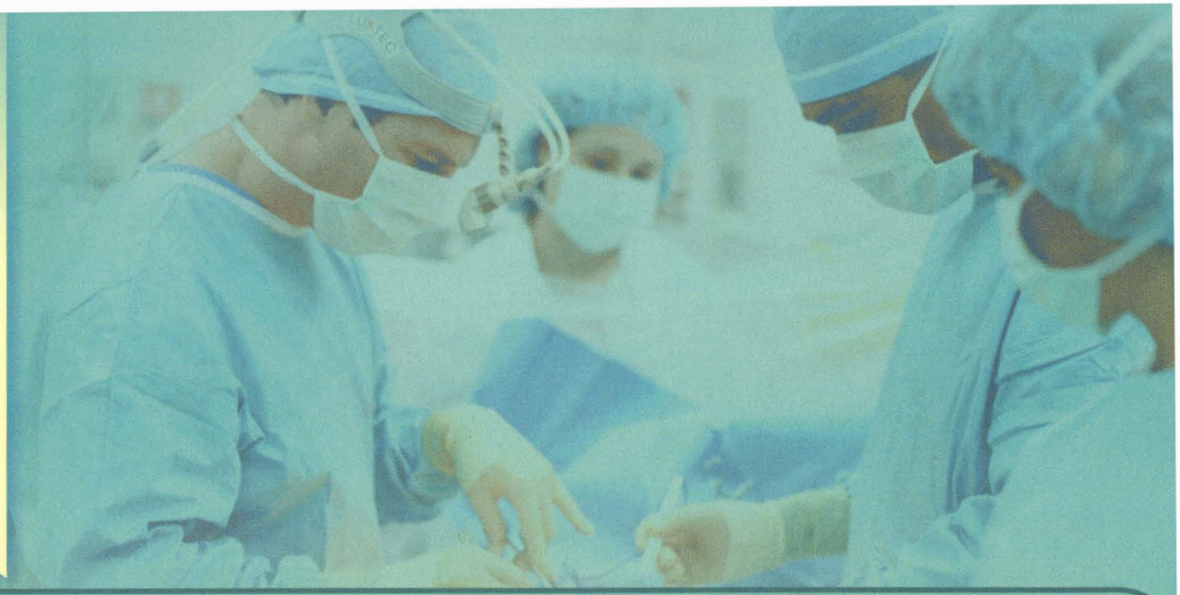
25. Which one of the following is not true in male breast cancer?
A. Most commonly it is infiltrating duct carcinoma
B. Mostly it presents as bleeding per nipple
C. Mostly mastectomy is required
D. Gynaecomastia is a predisposing factor

ANSWERS

1 C	2 C	3 D	4 B	5 C	6 D	7 A	8 C	9 A	10 D
11 B	12 C	13 C	14 D	15 A	16 B	17 B	18 D	19 A	20 D
21 D	22 D	23 D	24 D	25 B					

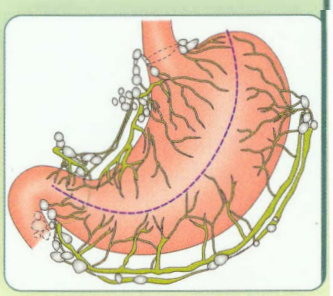
Section

II



Gastrointestinal Surgery

22. Oesophagus and Diaphragm
23. Stomach and Duodenum
24. Liver
25. Gall Bladder and Pancreas
26. Spleen
27. Peritoneum, Peritoneal Cavity, Mesentery and Retroperitoneum
28. Small Intestine
29. Large Intestine
30. Intestinal Obstruction
31. Rectum and Anal Canal
32. Lower Gastrointestinal Bleeding
33. The Appendix
34. Hernia
35. Umbilicus and Abdominal Wall
36. Blunt Abdominal Trauma, War and Blast Injuries and Triage
37. Abdominal Mass



22

Oesophagus and Diaphragm

- Surgical anatomy
- Physiology
- GORD
- Motility disorders
- Achalasia cardia
- Nutcracker oesophagus
- Carcinoma
- Stricture
- Perforations
- Diverticulum
- Dysphagia
- Surgical anatomy of the diaphragm
- Diaphragmatic hernia
- Tracheo-oesophageal fistula
- What is new?/Recent advances

SURGICAL ANATOMY

- Oesophagus is 25 cm in length, extending from the cricopharyngeal sphincter to the cardio-oesophageal junction (Fig. 22.1).
- Cardio-oesophageal junction lies to the left of T11 vertebra. It is identified at endoscopy by a Z-line.
- It is a jagged line where the oesophageal mucosa changes to gastric mucosa.
- **Collar of Helvitius:** It is the site at which the circular muscles of oesophagus turns to oblique muscles of the stomach at the incisura.
- It runs in the posterior mediastinum as a continuation of pharynx. 2 cm of this tube lies below the diaphragm.
- **Physiological constrictions** (Table 22.1)

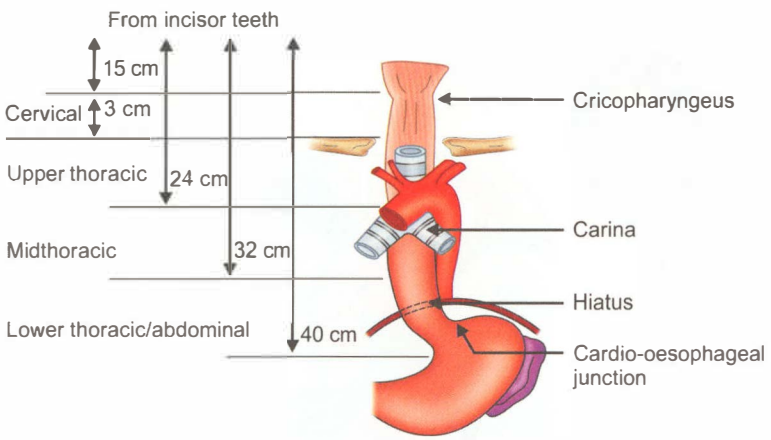


Fig. 22.1: Division of oesophagus into 4 parts based on distance from incisor teeth

Table 22.1 Physiological constrictions

Constrictions	Distance from incisor teeth	Diameter of oesophagus	Problems
1. Cricopharyngeal	15 cm	14 mm	Foreign body lodgement
2. Aortic and bronchial	25 cm	15–17 mm	Perforations during endoscopy
3. Diaphragmatic sphincter	40 cm	16–19 mm	Malignancy

Muscle layers (Figs 22.2 and 22.3)

- It has an inner circular layer and an outer longitudinal layer.
- Upper 1/3rd has striated muscle fibres.
- Lower 2/3rd has smooth muscle fibres.

PEARLS OF WISDOM

Clinically significant motility disorders affect lower two-thirds (smooth muscle) of oesophagus.

Mucosa

- The entire oesophagus is lined by squamous epithelium except last 3 cm which is lined by columnar cells. The columnar cells are similar to gastric mucosa but oxyntic and peptic cells are absent.
- **Mucosa is the toughest coat of oesophagus.**
- The smooth oesophageal mucosa gets transformed into rugal folds.

Lymphatic drainage (Key Box 22.1)

- Upper oesophagus drains into the left and right supraclavicular nodes.
- Middle oesophagus drains into the tracheobronchial nodes and paraoesophageal nodes.
- Lower oesophagus drains into lymph nodes along the lesser curvature of stomach and then into coeliac nodes. Involvement of coeliac nodes indicate inoperability.

Nerve supply

The parasympathetic nerve supply is by vagus nerve through extrinsic and intrinsic plexuses. The intrinsic plexus has **no Meissner's network** and Auerbach's plexus is present only in the lower two-thirds.

KEY BOX 22.1

LYMPHATIC DRAINAGE OF OESOPHAGUS

- Extensive lymphatic plexus in the submucosa which explains the easy spread
- Like in the colon they are classified as perioesophageal, paraoesophageal and lateral oesophageal lymph nodes
- Thus throughout the entire length of oesophagus various groups of lymph nodes are enlarged such as deep cervical, mediastinal, subcranial, hilar, diaphragmatic, para and peri-oesophageal, gastric, coeliac, etc.
- Presence of coeliac nodes indicate inoperability
- Longitudinal lymphatics are 6 times more than transverse. Hence, distant spread occurs fast—satellite nodules occur proximal and distal to the growth. This is the reason why 10 cm margin must be given in resection of the oesophagus
- One of the advantages of transhiatal resection over Ivor Lewis operation (*see* page 449)

Blood Supply (Figs 22.4 and 22.5)

Arterial

- **Cervical oesophagus:** Mainly from branches of the inferior thyroid artery.
- **Upper thoracic oesophagus:** Mainly from branches of the inferior thyroid artery and less consistently from anterior oesophagotracheal branch from aorta.
- **Mid and lower thoracic oesophagus:** Supplied by bronchial arteries.
- **Lower oesophagus:** Small branches of the left gastric artery.

Rich internal arterial anastomosis is present in the oesophagus and in the stomach. Hence, extensive mobilisation of oesophagus can be done without compromising viability.

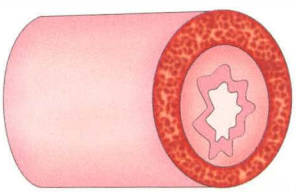


Fig. 22.2: You can see the multilayered structure. Endosonography detects all these layers

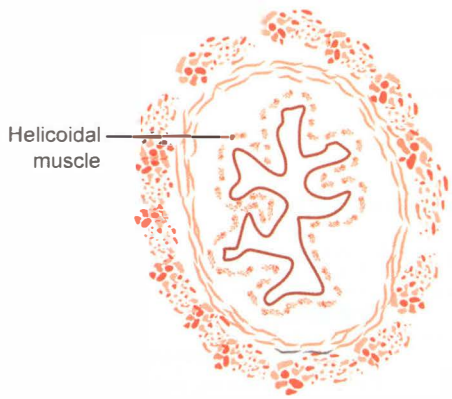


Fig. 22.3: Helicoidal muscle

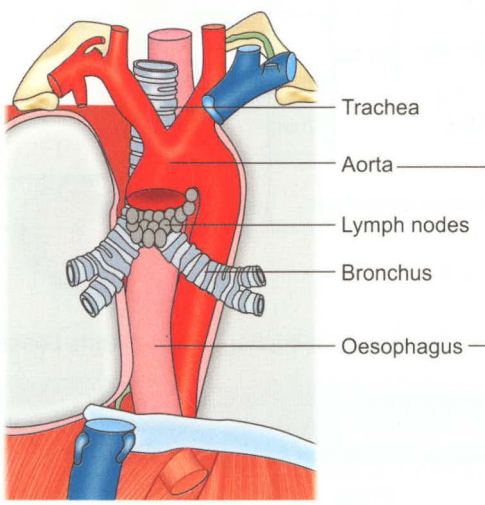


Fig. 22.4: Oesophageal anatomy—relationships

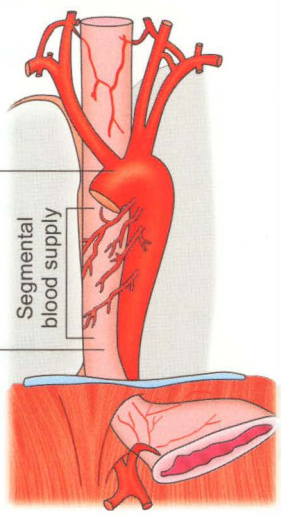


Fig. 22.5: Oesophageal anatomy—segmental blood supply

Table 22.2 Surgical anatomy of the oesophagus—key points

Surgical anatomy	Significance	Surgical points of interest
1. Upper oesophageal sphincter	Dense cricopharyngeus muscle	Zenker's diverticulum through Killian triangle
2. Lower oesophageal sphincter	High pressure zone at gastro-oesophageal junction	Weakness causes reflux oesophagitis
3. Oesophageal mucosa	Toughest coat of oesophagus	Important in oesophageal anastomosis
4. No serosa, no mesentery	Requires mobilisation	Leak rate is high, easy spread of carcinoma
5. Helicoidal muscle	Helps in peristalsis but it recoils due to elasticity	Hence, once resection takes place—specimen 'shortens' or retracts—anastomosis may become difficult especially in abdominal anastomosis
6. Segmental arterial supply	Extensive mobilisation can be done without compromising blood supply	Hence, transhiatal esophagectomy (THE) is done with ease
7. Lower end of oesophagus—veins	Rich intercommunicating veins between portal and systemic veins	Oesophageal varices occur here
8. Azygos vein crossing oesophagus in thorax	Can get injured and bleed	Resection of midoesophageal tumours
9. Mucosal and submucosal lymphatics out number capillaries	Oesophageal tumours extend over a long distance within oesophageal wall	Need for total oesophagectomy and cervical anastomosis + lymph node clearance

Venous drainage

Veins accompany corresponding arteries. Importantly, thoracic oesophagus drains into azygos and hemiazygos veins.

• Table 22.2

PHYSIOLOGY (Fig. 22.6)

The main function of the oesophagus is to transfer food from the mouth to the stomach. Voluntary contraction of the oropharynx pushes food into the upper oesophagus through a relaxed cricopharyngeal sphincter (Key Box 22.2). Then, due to primary and secondary peristalsis, the food bolus is transferred to the stomach.

KEY BOX 22.2

THE PHARYNGEAL STAGE—REFLEX

- Food stimulates mechanoreceptors
- Soft palate is pulled upwards so that food will not enter nasal cavities
- Vocal cords adduct and larynx is pulled upwards against epiglottis so that food will not enter trachea
- Cricopharyngeal sphincter opens
- Muscular wall of the pharynx then contracts from above downwards.

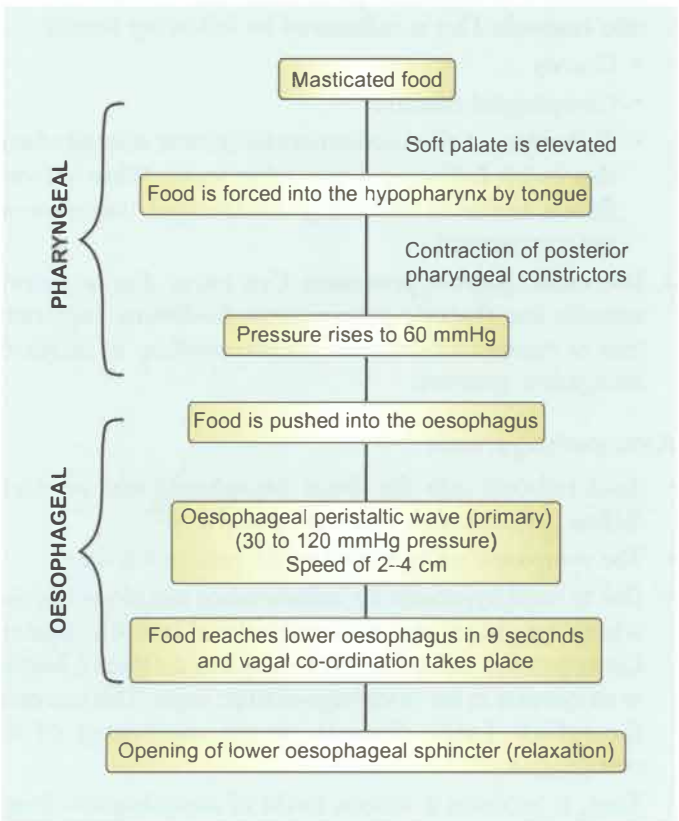


Fig. 22.6: Mechanism of swallowing—2 phases

- **Cricopharyngeal sphincter** is closed at rest. It helps in preventing regurgitation of oesophageal contents into the respiratory passage.
- **Lower oesophageal sphincter (LOS):** It is a physiological sphincter at the gastro-oesophageal junction. It is 3–4 cm long with a resting pressure of 20–25 mmHg. Hence, it is known as **high pressure zone (HPZ)**.
- This HPZ prevents reflux of gastric contents into the oesophagus. Cigarette smoking affects this zone and so, smokers have a high incidence of reflux oesophagitis.

GASTRO-OESOPHAGEAL REFLUX DISEASE

- Loss of competence of LOS leads to gastro-oesophageal reflux disease (GORD). The competence of LOS can be affected by obesity, smoking, excessive eating, etc. Sliding hernia is associated with GORD.
- As a result of reflux of gastric acid, extensive inflammation of the lower oesophagus occurs which results in various forms of oesophagitis.

Types

1. **Acute:** Following alcohol, burns, stress.
2. **Chronic:** It is associated with hiatus hernia or following oesophagojejunostomy.

It is mainly precipitated by

1. Structurally defective LOS secondary to inflammatory injury.
2. Inadequate abdominal length (short length) of oesophagus—precipitates a reflux when gastric distension occurs.
3. Ineffective oesophageal pump affecting clearance of food into stomach. This is influenced by following factors:
   - Gravity
   - Oesophageal motility
   - *Salivation*—Saliva neutralises the minute amount of acid that is left following a peristaltic wave. When salivary flow is decreased (following radiotherapy), the reflux can get exaggerated.
4. **Increased gastric pressure:** Can occur due to pyloric stenosis and diabetic gastroparesis. Following vagotomy, loss of receptive relaxation occurs resulting in increased intragastric pressure.

Aetiopathogenesis

- Acid refluxes into the lower oesophagus and produces diffuse inflammation with multiple ulcers.
- The symptoms are worse when the patient lies down.
- Due to vagal hyperactivity, inflammation and ulcers develop which produce severe longitudinal muscle spasm. Consequently, the cardia is drawn up into the thorax, leading to an increase in the oesophago-cardiac angle. This increases the reflux. Later, fibrosis causes shortening of the oesophagus.
- Thus, it becomes a vicious circle of *oesophagitis—longitudinal muscle spasm—displacement of oesophagus—increased regurgitation.*

Clinical features

Most common presentation of GORD is history of heart burn and regurgitation. Heartburn is confined to epigastrium and retrosternal areas, does not radiate to the back. Clinical presentation of the GORD can be classified as follows: Atypical symptoms can be so many and can thus confuse the picture.

Oesophageal	:	Dysphagia, regurgitation, heartburn
Gastric	:	Early satiety, belching, bloating, nausea
Pulmonary	:	Asthma, wheezing, aspiration, cough, dyspnoea, bronchitis, hoarseness of voice due to damage to vocal cords, etc.
Ear, nose, throat	:	Waterbrash, globus, hoarseness
Cardiac	:	Chest pain

What is globus? Sensation of a substernal lump (globus) When this occurs during fasting, it is termed 'globus hystericus'. It is a neurotic symptom in patients with emotional instability.

- **Retrosternal pain:** It is burning in nature and becomes worse on lying down. The pain reduces in the sitting position. The pain is described as *heart burn and can be confused for angina pectoris.* It is relieved on taking antacids.
- **Heart burn** is otherwise called **pyrosis**.
- **Occult blood in stools** and streaks of blood in the vomitus are common.
- **Anaemia** and weakness are uncommon features.
- **Dysphagia:** Transient difficulty in swallowing results from spasm due to inflammation of the lower end of oesophagus. Late dysphagia is due to stenosis or stricture of the oesophagus. Belching is not uncommon.

Johnson-DeMeester's scoring system

- Three important symptoms are taken into consideration

Grade	*Features*
	Heart burn
0	None
1	Minimal—occasional episodes
2	Moderate—medical therapy visits
3	Severe—interferes with daily activities
	Regurgitation
0	None
1	Minimal—occasional episodes
2	Moderate—on position or straining
3	Severe—features of aspiration
	Dysphagia
0	None
1	Minimal
2	Needs fluid to clear
3	Causes food impaction

Modified Savary Müller classification of reflux oesophagitis

Grade I: Single or isolated erosion at or above GE junction

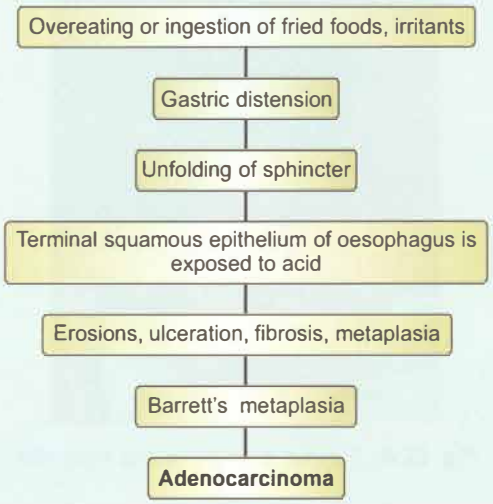
Grade II: Multiple non-circumferential erosions above GE junction

Grade III: Circumferential erosions above GE junction

Grade IV: Chronic lesion—stricture, ulceration/short oesophagus

Grade V: Columnar epithelium in continuity with Z line (Barrett's oesophagus)

GORD—SUMMARY



Investigations (Table 22.3, Key Boxes 22.3 and 22.4)

- **Barium swallow**[1] in the Trendelenburg's position (head down position) can demonstrate the reverse flow of barium into the lower end of the oesophagus (from the stomach).
- **Oesophagoscopy** may reveal red, angry looking mucosa in the lower end of the oesophagus.
- **Oesophageal manometry** to detect motility disorders.
- 24-hour pH monitoring is the gold standard.

KEY BOX 22.3

OESOPHAGEAL MANOMETRY

- Information about LOS
  - Resting pressure
  - Length
  - Relaxation
- Quality of oesophageal peristalsis
- Manometry is essential. To place the pH probe 5 cm above upper border of LOS for ambulatory pH monitoring.

KEY BOX 22.4

24-HOUR pH MONITORING IS THE GOLD STANDARD

- Indications: When symptoms are certain but endoscopy is normal, atypical symptoms
- It involves the transnasal placement of a pH measuring electrode in the lower oesophagus. The pH electrode monitors the changes in intra-oesophageal pH over 24 h
- A 24 h pH profile is thus obtained that provides information on frequency, duration and pattern of reflux
- A reflux episode is defined as a pH drop to below pH 4
- Thus identification, type and duration of reflux is noted

Medical management

Treatment of uncomplicated GORD can be discussed under following headings.

I. Lifestyle modification

- Stop smoking
- Stop alcohol
- Control obesity
- Avoid coffee, chocolate and coke
- Head up—propped-up position
- Avoid stooping
- Avoid tight garments

II. Drugs

- Antacids with alginate—antireflux floating alginate.
- **Proton pump inhibitors:** Pantoprazole 40 mg, esomeprazole till 20 mg may have to be given for one or two months or full symptoms are controlled. These are antisecretory drugs.
- **Prokinetics:** Itopride 50 mg can be given 2–3 times a day for 8 weeks on empty stomach. Prokinetics enhance motility. Cisapride and mosapride are not favoured because they can cause cardiac arrhythmias.

III. Mucosa protective agents

- Sucralfate colloidal bismuth---cytoprotective agent.
- It is a sucrose sulfate–aluminium complex which binds to the mucosa. Thus it protects mucosa of GI tract against hydrochloric acid.
- Colloidal bismuth compounds.

Table 22.3 Investigations of GORD

To detect structural abnormalities	To detect increased exposure to gastric juice	To detect functional abnormalities
A. Barium swallow • Detects hiatus hernia (prone position) • Detects stricture or any rings • Extrinsic compression can be detected • Can also detect small carcinoma ***B. Flexible endoscopy*** • Can detect hiatus hernia or diverticulum, etc. • Can detect oesophageal abnormalities	***A. Flexible endoscopy and to biopsy the Barrett's oesophagus*** ***B. Grading of oesophagitis by endoscopy*** Grade I: Small, circular, nonconfluent erosions Grade II: Linear erosions lined with granulation tissue that bleeds on touch Grade III: Erosions with circumferential loss of the epithelium---cobblestone oesophagus Grade IV: Stricture	***A. Oesophagus manometry*** • It is indicated when motor abnormality is suspected such as achalasia, diffuse spasm • Can also detect scleroderma polymyositis • Average LOS pressure < 6 mmHg, suspect GORD • High resolution manometry is more accurate

[1] Barium swallow: Thick paste to see the oesophagus. Barium meal: A thin paste to see the stomach. Both are radio-opaque.

IV: Endotherapy

- Endoscopic plication/suturing
- Enteryx injection
- **Plexiglass microspheres (PMMA):** Through an endoscopic needle, microspheres suspended in gelatin are injected. Gelatin is absorbed and spheres cause tissue bulking.

KEY BOX 22.5

MEDICAL MANAGEMENT OF GORD

- **A**lcohol to be minimised
- **L**ose weight
- **C**offee/tea to be minimised
- **O**esophageal mucosa protectors—antacids, H_2 receptor blockers
- **H**ead—up tilt
- **O**ily and spicy food must be avoided.
- **L**arge meal to be avoided at night times

Remember as **ALCOHOL**

V. Surgery: Indication

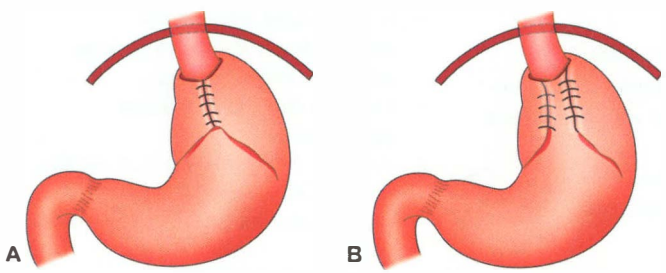
To summarise, quit smoking, decrease alcohol intake, avoid overweight and start walking. Do not drink coke and do not eat chocolates, take mucosa protective agents (Key Box 22.5).

1. Intractable pain
2. Complications such as haemorrhage or stricture
   - The results of antireflux surgery are good with a small mortality rate (0.1 to 0.5%). However, careful selection of patients depending upon their symptoms and lifestyle are important factors.

Types of surgery

1. Nissen's total fundoplication (Figs 22.7A and B): The aim is to restore 2–4 cm of intra-abdominal oesophagus by reducing the hernia, followed by repair of the hiatus.

- In this operation, fundus of the stomach is mobilised by dividing short gastric arteries.



Figs 22.7A and B: Nissen's fundoplication

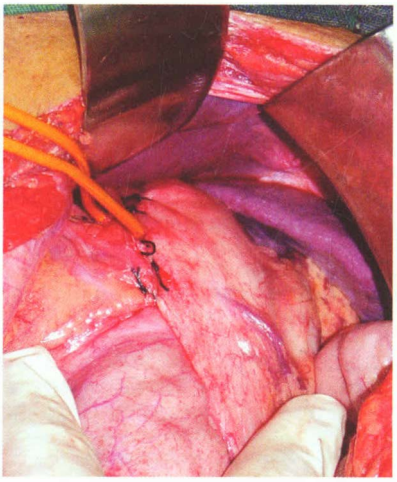


Fig. 22.8: Fundus brought on the right side

- Fundus is brought behind the oesophagus and wrapped in front of oesophagus (Fig. 22.8). It is a ***loose wrap (Floppy Nissen's).***
- Diaphragmatic defect is repaired by using nonabsorbable sutures such as nylon or silk. This is the operation which serves all aims.
- Mortality and morbidity should be minimised.

Laparoscopic fundoplication

- Most popular today
- Minimal morbidity and mortality
- Early discharge, within 1–2 days
- Early recovery
- All operative steps that are performed in open surgery are carried out here with a 'better vision' in laparoscopic method.

Principles of fundoplication

- 360° gastric fundoplication should be no longer than 2 cm.
- It should be **constructed over a 60 F bougie**.
- Fundoplication should be placed in the abdomen without tension.
- **Only 'fundus'** should be used to wrap (fundus relaxes—body does not relax on swallowing).
- **Vagus should not be damaged** because it may result in failure of sphincter to relax.
- Lengthen the oesophagus with a **Collis gastroplasty** (in cases of short oesophagus).
- Patients with **normal peristaltic contractions** do well with 360° Nissen's fundoplication—for others, two-thirds partial fundoplication may be the procedure of choice.

Complications

- Too tight a plication may result in dysphagia or gas bloat syndrome wherein belching is prevented.

2. Partial fundoplication (Toupet) solves the above problem wherein fundus is sutured around the back of oesophagus (Fig. 22.9) or Dorr's, where fundus is sutured anterior to the oesophagus.

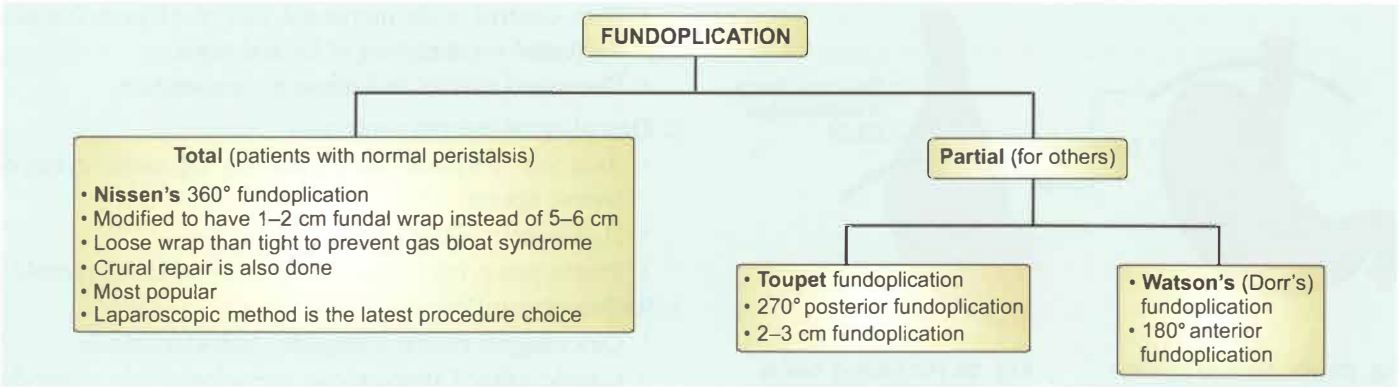


Fig. 22.9: Types of fundoplication

3. Belsey Mark IV operation

- There are 3 layers of sutures to be placed in this operation *via* a thoracotomy (Key Box 22.6).
- **First row**: Four interrupted silk mattress sutures are placed between the anterior surface of the oesophagus and adjacent fundus of the stomach so as to wrap the stomach around the anterior two-thirds of the oesophagus.
- **Second row:** Sutures are placed between oesophago-gastric junction and under surface of the diaphragm to maintain the junction below the diaphragm.
- **Third row:** Posterior crural sutures are placed to tighten the opening.

KEY BOX 22.6

ADVANTAGES OF BELSEY MARK IV

- Reduces size of the ring
- Creates a valve at the oesophagogastric junction in order to prevent reflux
- Recurrence is low

4. Hill's repair: Median arcuate ligament repair.

- In this, the long intra-abdominal segment of the oesophagus is firmly fixed below the diaphragm by anchoring the gastro-oesophageal junction to the crura just above the **median arcuate ligament**. This is described as **posterior gastropexy**.

5. Recurrent GORD: Success of revision surgery is much less than primary surgery. Partial gastrectomy with **Roux-en-y** reconstruction is the final surgery for GORD. It diverts bile and pancreatic juice and reduces acid secretion.

RECENT ADVANCES

Endoscopic treatment of GORD: 3 types

1. Radiofrequency energy to LES: It changes sphincter compliance
2. Barrier at GE junction: An ethylene vinyl alcohol copolymer is injected into submucosa 1–2 mm caudal to Z-line.
3. Direct endoscopic suturing and tightening of LES.

HIATUS HERNIA

Definition

Abnormal protrusion of abdominal viscus through the oesophageal hiatus into the chest.

Types (Key Box 22.7)

1. **Sliding hernia or oesophagogastric hernia:** It is the commonest type of hiatus hernia, accounting for about 80% of cases (Fig. 22.11). It may be associated with GORD.
2. **Rolling or para-oesophageal hernia**
3. **Mixed hernia**
4. **Massive herniation**

Common symptoms

1. **Symptoms due to reflux:** Regurgitation and heart burn are the two most common symptoms.
2. **Symptoms due to complications:** They are dysphagia, odynophagia, haematemesis and melaena.
3. **Nonoesophageal symptoms:** They are asthma and chest pain.

SLIDING HERNIA (Figs 22.10 and 22.11)

Anatomical factors which prevent sliding hernia

1. Presence of 2 cm of intra-abdominal oesophagus.
2. **The angle of His:** The oesophago-cardiac angle of about 45° has valvular effect.
3. Mucosal folds at the oesophago-cardiac junction.
4. Positive intra-abdominal pressure which closes the abdominal oesophagus.

KEY BOX 22.7

TYPES

I	: H^0	:	Normal
II	: H^1	:	Sliding
III	: H^2	:	Para-oesophageal
IV	: H^3	:	Mixed
V	: H^4	:	Massive herniation

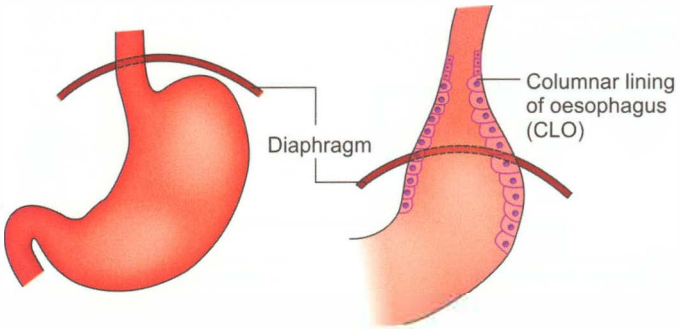


Fig. 22.10: Normal position

Fig. 22.11: Sliding hernia

5. **Lower oesophageal sphincter (LOS):** It is a functional sphincter which increases the pressure during coughing, straining, etc.

Causes of sliding hernia[1]

1. Deposition of **fatty tissue** in the hiatus leads to weakening of the hiatus in obese individuals.
2. Advancing **age** resulting in muscular degeneration can predispose to hernia.
3. Raised intra-abdominal pressure due to lower **abdominal tumours**, pregnancy, etc.
4. **Saint's triad:** Gall stones, diverticulosis and hiatus hernia can occur together in a patient.

Clinical features

- Sliding hernia produces symptoms like reflux oesophagitis.
- More common in women, especially if obese.

Investigations

- **Oesophagoscopy** reveals varying degree of inflammation. During oesophagoscopy, when the patient is asked to strain (Valsalva's manoeuvre), the sphincter is seen to be more patulous and herniation of gastric mucosal folds can be seen. **Reflux of the gastric acid is the most valuable sign.**
- **Barium meal** demonstrates gastro-oesophageal reflux in the Trendelenburg position.

Treatment

I. Conservative treatment

In all cases of GORD, conservative treatment has to be tried first. The results of surgery are appreciated only when the symptoms are significant and conservative treatment fails.

PEARLS OF WISDOM

Oh LORD, save me from GORD!

Principles

1. Lifestyle changes
   - Decrease in weight
   - Diet control with increased intake of proteins and decreased consumption of fat and sugar.
   - Decreased alcohol and tobacco consumption.
2. Oesophageal mucosa protection
   - *Antacids:* Preparations containing alginates, cytoprotective agents.
   - H_2 blockers: Ranitidine.
   - Proton pump inhibitors: Omeprazole or esomeprazole.
3. **Reflux prevention**
   - Oesophageal reflux: Cisapride, metoclopramide
   - Gastric reflux: Domperidone, metoclopramide, cisapride.
4. Decision of surgery.

ROLLING HERNIA

In this condition, cardio-oesophageal junction is normal. Greater curvature of the stomach ascends into a preformed sac in the mediastinum (Fig. 22.12). Thus, there are no features of reflux oesophagitis but the sac containing stomach in the thorax causes compression of the heart and lung.

Clinical features

- No retrosternal burning pain because no reflux
- Discomfort after a small meal
- Feeling of fullness after a meal or dysphagia due to large sac
- Palpitations due to compression on the heart
- Respiratory tract infection and hiccough due to irritation of phrenic nerve.

Investigation

Barium meal shows the sac in the thorax containing stomach. Sometimes, it can be upside down.

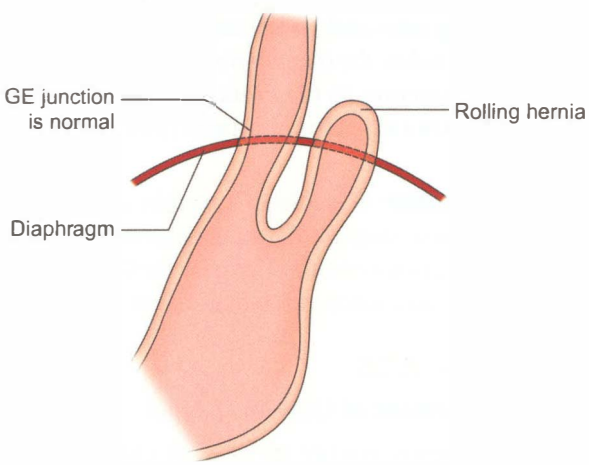


Fig. 22.12: Rolling hernia

[1]Causes can be remembered as FATTY (F: fat, A: age, T: tumour, T: triad (Saint's triad), Y: yielding hiatus.)

Treatment

Reduction of the sac and repair of the hiatus by using nonabsorbable suture material to approximate the right crus of the diaphragm.

MIXED HERNIA

- In this, both sliding and rolling hernia are present (Fig. 22.13).
- Symptoms are mixed.
- Treatment is also mixed and is done for both sliding and rolling hernia.

Complications of GORD

1. **Stricture oesophagus:** It is seen in middle-aged and elderly patients. Due to repeated reflux, ulcers, fibrosis and stricture develop in the lower end of the oesophagus. Early diagnosis by endoscopy followed by **frequent dilatation** and **proton pump inhibitors** will help the situation.
   - Peptic strictures are difficult to manage surgically.
   - Surgery is indicated in refractory cases of dilatation, in the form of **gastroplasty**.

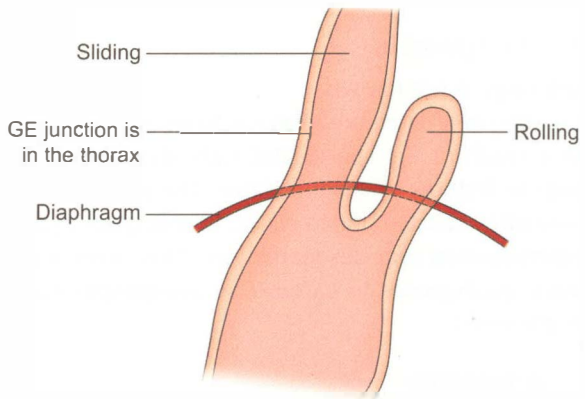


Fig. 22.13: Mixed hernia

2. **Oesophageal shortening** is also treated by Collis gastroplasty by using stomach (short oesophagus).
3. **Barrett's oesophagus:** Also known as columnar-lined oesophagus (**CLO**) (details are given below).

BARRETT'S OESOPHAGUS

Definition

- When columnar mucosa extends at least 3 cm into oesophagus or when it shows intestinal metaplasia, it is described as Barrett's oesophagus (Figs 22.14 to 22.17).
- Ulcer in the Barrett's CLO is called **Barrett's ulcer**.

Pathogenesis

Repeated reflux results in shifting of the oesophago-gastric junction upwards, which further increases the reflux resulting in intestinal metaplasia of **middle and lower oesophagus.**

Pathological types

1. **Gastric type** with chief and parietal cells.
2. **Intestinal type** with ***goblet cells*** is a marker of intestinal metaplasia. This mucosa is smooth (unlike gastric folds).
3. **Junctional type:** It has mucus glands and resembles gastric cardia.

Clinical types

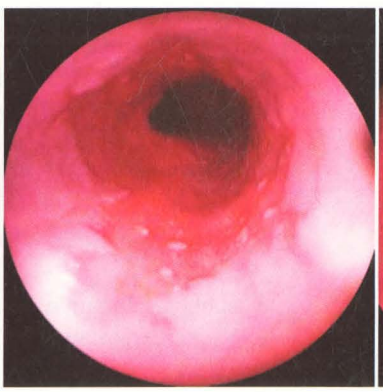
1. **Long segment:** Metaplastic changes involving more than 3 cm.
2. **Short segment:** Metaplastic changes involving less than 3 cm.

Incidence of malignancy

40 times more prone to carcinoma of the **lower and middle oesophagus** as compared to the general population.

Types of dysplasia

Low grade: Negligible risk of carcinoma



Figs 22.14 and 22.15: Endoscopic view of reflux oesophagitis and Barrett's oesophagus

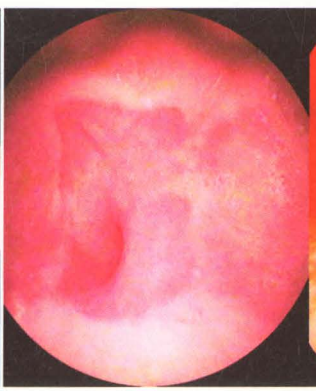


Fig. 22.16: Barrett's oesophagus with stricture

Fig. 22.17: Carcinoma GE junction in Barrett's oesophagus

KEY BOX 22.8

RISK FACTORS FOR CARCINOMA

- CLO > 8 cm
- Smoking
- Reflux due to previous gastric surgery
- High-grade dysplasia—indications for oesophagectomy

- High grade: Very high-risk of carcinoma (Key Box 22.8)

Screening programme

- It is important to screen these patients regularly with **repeat endoscopies and multiple biopsies to find dysplasia** (Key Box 22.9).

KEY BOX 22.9

CARCINOMA IN BARRETT'S OESOPHAGUS

- It will be invasive
- It is more proximal
- Carries poor prognosis
- 40 times increased risk compared to general population

Treatment

- High dose proton pump inhibitors for 8 weeks.
- Laser photodynamic therapy.
- Argon beam plasma coagulation
- Asymptomatic, symptomatic Barrett's oesophagus responds well to laparoscopic antireflux surgery. Antireflux surgery also prevents progression to high-grade dysplasia or adenocarcinoma.
- Endoscopic mucosal resection for Barrett's oesophagus with low-grade dysplasia.
- Oesophagectomy in cases of high-grade dysplasia.

PEARLS OF WISDOM

Antireflux surgery may protect mucosa against development of dysplasia in patients with Barrett's oesophagus.

Complications of Barrett's oesophagus

1. **Oesophageal ulcers:** Barrett's ulcer—pain, bleeding and perforation.
2. **Oesophageal stricture:** Usually located in the middle or upper oesophagus.
   - **Peptic stricture occurs in the distal oesophagus.**
3. **Dysplasia and adenocarcinoma.**

Non-reflux Oesophagitis

This is also a condition which occurs due to several factors such as corrosives, drugs, chemoradiation, AIDS, etc. They are summarised in Table 22.4. Basic treatment is similar to that of reflux oesophagitis and avoid the causative agents.

Table 22.4 Types of non-reflux oesophagitis

	Agents causing	Clinical features
1. Corrosive:	Lye, acid, alkali	Burns, stricture
2. Infective:	Candida, Herpesvirus	Chronic illness, AIDS
3. Radiation:	20–40 Gy units to mediastinum	Ulceration and stricture
4. Chemotherapy	Adriamycin	—
5. Drug induced:	NSAID, doxycycline	↑ reflux

MOTILITY DISORDERS OF THE PHARYNX AND OESOPHAGUS

PLUMMER-VINSON SYNDROME (Key Box 22.10)

It is a precancerous condition in which there is a severe spasm of circular muscle fibres at the cricopharyngeal sphincter level or pharyngo-oesophageal junction, and it is associated with **development of postcricoid web.**

Aetiopathogenesis

- Aetiology is not known
- It is seen in women who have worries and anxiety.
- As a result of the spasm and web, dysplasia occurs and leads to features of anaemia later. The proximal mucosa is constantly irritated due to stasis. It undergoes hypertrophy, hyperkeratosis and desquamation. This, over a period of years, predisposes to carcinoma oesophagus (carcinoma oropharynx).

Clinical features

- Women in the middle age group are affected.
- Increasing **dysphagia** for solids and liquids due to spasm.
- Features of **anaemia**—pallor, stomatitis, ulcerations, bald tongue (without papillae), koilonychia, splenomegaly and microcytic hypochromic anaemia.
- As a result of obstruction, the fluid tends to spill over into the larynx giving rise to recurrent aspiration, respiratory tract infection, cyanosis or choking.

KEY BOX 22.10

PLUMMER-VINSON SYNDROME

- Women
- Dysphagia, anaemia
- Cricopharyngeal spasm
- Dilatation
- Iron supplements
- Precancerous condition

Treatment

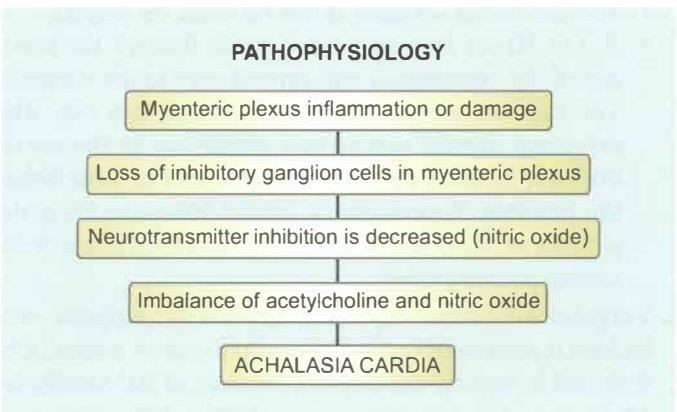
- Reassurance
- Improving anaemia: Iron tablets or blood transfusion and correction of nutritional deficiencies.
- Regular dilatation by using gum elastic bougies.

ACHALASIA CARDIA

- It is a primary oesophageal motility disorder.
- It is also called cardiospasm because of severe spasm of the circular muscle fibres of the lower end of the oesophagus. The contracted segment does not relax during the act of swallowing (achalasia = failure of relaxation). As a result of this, there is dilatation, tortuosity and hypertrophy of the oesophagus above.
- Incidence: 6 in 1,00,000 population/year.

Aetiopathogenesis

1. **Idiopathic:** This occurs due to absence or degeneration of Auerbach's plexus throughout the body of the oesophagus leading to improper integration of the parasympathetic impulse. This is called primary achalasia.
2. **Acquired variety** is seen in South American countries caused by *Trypanosoma cruzi*[1] (sleeping sickness)—**Chagas disease**. This organism destroys the ganglion cells of the Auerbach's plexus.
3. Stress and emotional factors and vitamin deficiencies are also associated with this disease.
   See chart below for pathophysiology.



- Classical achalasia: Loss of ganglion cells and neural fibrosis occurs.
- Vigorous achalasia: No loss of ganglion cells but ganglionitis occurs.

Clinical features

- Women around 30–40 years of age are commonly affected. The ratio of affected females to males is 3:2.
- **Dysphagia** develops slowly and it is progressive.
  - Solids, by forming a bolus and aided by gravity, as they touch the contracted segment, may partially open up the sphincter. Thus, there is no dysphagia for solids. Dysphagia for liquids is an important feature and it results in **regurgitation** (oesophageal pseudovomiting). The regurgitant material contains foul-smelling oesophageal contents. Malnourishment, ill health and weight loss follow soon. **Dysphagia, regurgitation and weight loss form the triad of achalasia cardia.**
- **Recurrent respiratory tract infection** due to spillage of liquids can also occur.
- **Features of anaemia**—glossitis, stomatitis, pallor, bald tongue.
- **Retrosternal discomfort** and radiation of pain to the interscapular region may be present.
- **Pseudoachalasia:** Tumours of cardia mimicking achalasia. Often patients present with features of recent achalasia (dysphagia). During endoscopy, some difficulties are encountered and once the scope enters the GE junction, growth will be seen.

PEARLS OF WISDOM

Inhalation of amyl nitrite leads to sphincter relaxation in achalasia but not in pseudoachalasia.

Investigations

1. **Barium swallow**
   - Uniformly dilated oesophagus above, with a smooth tapering segment below—**cucumber oesophagus**.
   - In chronic cases, it may be sigmoid-shaped (Figs 22.18 and 22.19).
2. **X-ray chest**
   - Mediastinal mass (pseudotumour)[2] produced by dilated oesophagus can be seen (Fig. 22.20).
   - **Retrocardiac air-fluid level is seen in the lateral view.**
   - Aspiration pneumonitis can be diagnosed.
3. **Plain X-ray abdomen erect:** Fundic air bubble is absent because of the stasis of fluid in the oesophagus.
4. **Oesophagoscopy:** It reveals a dilated sac containing stagnant food and fluid due to stasis which splashes out with each heart beat and with each respiratory movement.

   LES: Lower oesophageal sphincter is closed, with air insufflation. It has a 'rosette' appearance (Fig. 22.21).
   - Oesophagoscopy is also done to rule out proximal malignancy.
   - Also done to evaluate oesophagitis, stricture or a tumour at cardia.

[1] *Trypanosoma cruzi* causes megaoesophagus, megaduodenum, megacolon and megaureter. Can we call it a 'mega'organism?

[2] Aortic aneurysm, cold abscess (paravertebral), scoliosis are the other pseudotumours in the chest X-ray.

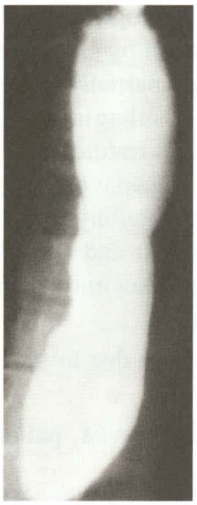


Fig. 22.18: Barium swallow showing dilated oesophagus

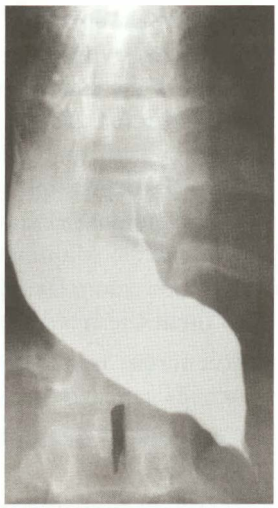


Fig. 22.19: Barium swallow showing sigmoid oesophagus

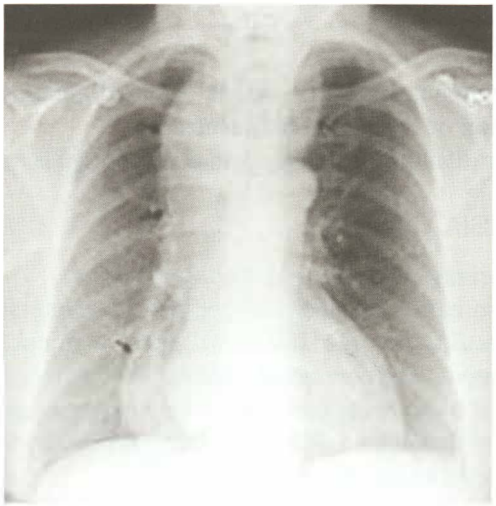


Fig. 22.20: Achalasia—chest X-ray

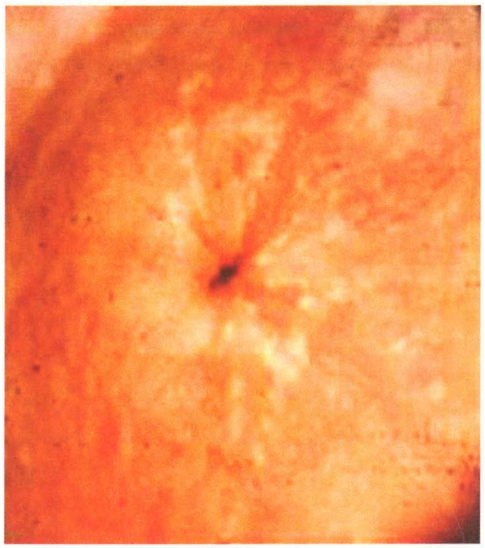


Fig. 22.21: Endoscopy showing rosette appearance

5. **Oesophageal manometry:** Following features are characteristic of achalasia cardia:
   - Hypertensive lower oesophageal sphincter (LOS): It does not relax on swallowing.
   - **Aperistalsis in the body** of oesophagus
   - Increased resting pressure in the oesophagus
6. **Ultrasound:** It may detect subepithelial tumour infiltration in secondary achalasia due to a distal carcinoma.

PEARLS OF WISDOM

You suspect of achalasia by clinical features
You support achalasia by Barium swallow
You confirm achalasia by manometry
You exclude carcinoma by UGI scopy
You treat by dilatation or surgery

Treatment (Table 22.5—four choices)

- Aim of the treatment is to cure the disease.
- **More importantly, the obstruction at the lower end o oesophagus must be relieved.**
- Modified laparoscopic Heller's cardiomyotomy is the choice of surgery now.

1. **Heller's cardiomyotomy:** The aim is to reduce outflow resistance at the lower oesophageal sphincter.
   - With a left thoracoabdominal incision, the oesophagus and the stomach are completely mobilised.
   - The contracted segment is felt between the fingers.
   - A 7 to 10 cm long incision is made through the lower end of the oesophagus and carried over to the stomach. The muscles are cut till the mucosa bulges out. **The myotomy should extend proximally up to the aortic arch and distally up to the stomach to 1 to 2 cm below the junction.** Success rate is around 90%. 3 to 5% of the patients develop reflux oesophagitis which needs to be treated conservatively.
2. **Forceful dilatation:** By using **Pneumatic balloon**—the balloon is positioned under fluoroscopic control within LOS. It should be rapidly inflated to a pressure of 300 mmHg for 15 seconds. Success rate is around 70%. 20% chances of reflux are present. Oesophageal perforation can also occur. Recurrences are common (Figs 22.22 and 22.23).
3. **Injection treatment:** **Inj. Botulinum toxin** is injected into the lower oesophageal sphincter (LOS) endoscopically. Injection acts by interfering with cholinergic excitatory neural activity at LOS and blocking acetylcholine release from nerve terminals. It is temporary and repeat injections are necessary.
4. **Drugs:** Sublingual nifedipine can produce short-term relief.
5. **Endoscopic myotomy—popularly called POEM:** In this, through endoscope, a submucosal injection of a mixture of indigo rouge, epinephrine, and saline is done. A 2 cm long

Table 22.5 Treatment of achalasia cardia

Pharmacological therapy	Botulinum toxin injection	Pneumatic dilatation	Surgery
• Sublingually used	• BoTx A 80–100 units are injected in one sitting	• Most effective nonsurgical option	• Modified lap. Heller's cardiomyotomy is the gold standard
• Nitrates and calcium channel blockers	• 20–25 IU are injected into each quadrant	• It disrupts the circular muscle fibres of LES	• 6 cm of lower oesophagus and 1 to 2 cm of proximal stomach myotomy is done
• Nitrate 5–20 mg before meal and Ca channel blockers 10–30 mg before meal	• Use 5 mm sclerotherapy needle	• Polyethylene balloon is used	• 90% success
• They relax smooth muscle	• It acts by blocking acetylcholine release	• Dilators are 3, 3.5 and 4 cm	• 3% reflux oesophagitis
• Nitrates have more intense effect and more side-effect vice versa with Ca channel blocker	• Repeat injections are necessary as effect is temporary	• 70% permanent relief	
• Headache is the side-effect	• Pregnancy, hypersensitivity to albumin are contraindications	• Side-effect is perforation, gastro-oesophageal reflux, haemorrhage	

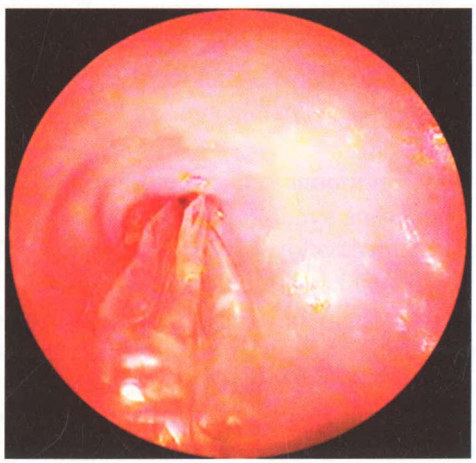


Fig. 22.22: Oesophageal balloon dilators

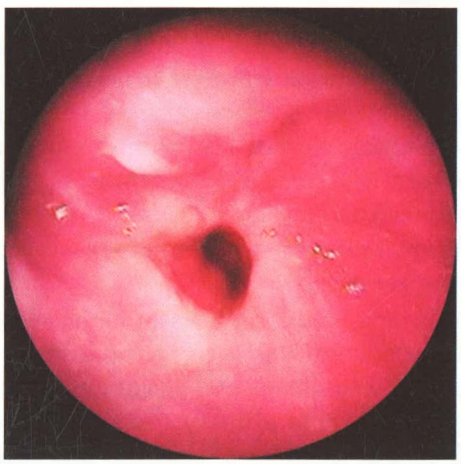


Fig. 22.23: Oesophageal narrowing of lower end

incision is made and submucosal tunnel is created. Then circular muscle bundle is cut slowly, beginning from 7–8 cm above GE junction to 2–3 cm below GE junction.

Complication of achalasia cardia

- Due to prolonged stasis and chronic irritation, it can predispose to **carcinoma** of the mid and lower oesophagus (due to metaplasia). Hence, it is a precancerous condition.
- Squamous cell carcinoma is the most common type identified in a patient with achalasia.

DIFFUSE OESOPHAGEAL SPASM

An entity of unknown aetiology, presents with dysphagia and chest pain.

Aetiology: Exact cause is not known. *Stress* may be one of the factors (Fig. 22.23).

Symptoms and signs

Discomfort in the chest (chest pain) or spasmodic pain, dysphagia, weight loss.

Investigations

1. **Barium swallow:** It is typically described as corkscrew oesophagus—segmental spasms, area of narrowing and irregular uncoordinated peristalsis.
2. **Manometry:** It is the diagnostic test. Findings are normal lower oesophageal sphincter, alteration of oesophageal peristalsis and contractions.
3. **Ambulatory 24-hour pH monitoring:** This is an important test to rule out GORD.

Treatment

It is directed in the similar lines of achalasia cardia—medical therapy, pneumatic dilatation intra-sphincteric botulinum toxin injection, Heller's myotomy and partial fundoplication.

NUTCRACKER OESOPHAGUS

- Aetiology is not known
- Chest pain is the most common symptom followed by dysphagia.
- Barium swallow is usually normal. An epinephric diverticulum is usually present.
- Majority of the patients have normal propagation of peristaltic waves. However, in the distal oesophagus, peristaltic waves have very high amplitude (>180 mmHg) and duration (> 6 seconds).
- Dilatation, myotomy, proton pump inhibitors, calcium channel blockers are used with limited success.

CARCINOMA OF THE OESOPHAGUS

Introduction

- Majority of oesophageal cancers are squamous cell carcinoma, worldwide. However, in most Western countries, more than 70% are adenocarcinomas. This shift is mainly due to smoking, alcohol and Barrett's oesophagus. Some improvement has taken place now because of early diagnosis, detection and multimodality treatment.
- **Oesophageal cancers have poor prognosis because of the following reasons:**
  - More lymph vessels are present in the submucosa than blood capillaries and hence, spread is fast.
  - Lymph flow in submucosal plexus runs in a longitudinal direction. Hence, primary tumour can extend for a considerable length both superiorly and inferiorly.
  - Cervical oesophagus has direct spread to regional nodes and there are less lymphatics in the submucosa. Hence, prognosis is better.

What is the 'change' in carcinoma oesophagus?

- Squamous cell carcinoma is more common in Africans and Asians. Adenocarcinoma is more common in whites and Americans. Adenocarcinoma is increasing.
- Survival is improving
- There is improvement in detection and staging.
- Detected more frequently in younger patients.
- Detected at an early stage also.

AETIOPATHOGENESIS

I. Precancerous/predisposing conditions

1. GORD with Barrett's oesophagus (*see* page 432)

PEARLS OF WISDOM

Malignancy in Barrett's oesophagus is curable if detected at an early stage. Hence, endoscopic surveillance is recommended.

2. Plummer-Vinson syndrome with squamous metaplasia (*see* page 438)—upper oesophagus.
3. Achalasia cardia (*see* page 439)—lower oesophagus.
4. Corrosive strictures (*see* page 451).
5. **Familial keratosis palmaris or plantaris (tylosis):** It is a condition inherited as an autosomal dominant trait. It has increased incidence of oesophageal cancer.

II. Carcinogens

1. Tobacco, heavy smoking and abuse of alcohol increases incidence of both squamous cell carcinoma and adenocarcinoma by almost 20–25%.
2. **Human papilloma virus (HPV) infection increases squamous cell carcinoma.** This oncogenic virus, has been associated with cervical and oropharyngeal cancers also.
3. **Dietary carcinogens and squamous cell carcinoma**
   - A. Elevated nitrates in drinking water.
   - B. Food (pickles, corn) containing fungi—*Geotrichum candidum* as in endemic areas of China.
   - C. Baked bread
4. **Factors for squamous cell carcinoma**
   - A. Hovels-Evans syndrome or tylosis—40 to 60% of the patient develop cancers. Also called familial keratosis plantaris.
     Tylosis: (focal nonepidermolytic palmoplantar keratoderma) is a rare disease inherited in an autosomal dominant manner that is characterised by hyperkeratosis of the palms and soles and oesophageal papillomas. Patients with this condition exhibit abnormal maturation of squamous cells. They also cause inflammation within the oesophagus and are at extremely high risk of developing oesophageal cancer.
   - B. **Lye strictures up to 30%:** Squamous cell carcinomas may arise in lye strictures, often developing 40 to 50 years after caustic injury. The majority of these cancers are located in the middle third of the oesophagus.
   - C. Achalasia—30%
   - D. Oesophageal web—20%
   - E. Plummer-Vinson syndrome —10%
   - F. Short oesophagus
   - G. Peptic oesophagitis
   - H. Patients with head and neck cancers—field cancerisation
   - I. Patients with coeliac disease
   - J. Drinking boiling hot tea/coffee

5. **Factors for adenocarcinoma**
   A. Barrett's oesophagus
   B. **Obesity:** Increased body mass index is a risk factor for adenocarcinoma of the oesophagus, and individuals with the highest body mass index have up to a seven-fold greater risk of oesophageal cancer than those with a low body mass index. Probably, the linkage between obesity and gastro-oesophageal reflux disease is presumed to be a chief factor.
   C. Reflux oesophagitis
6. Low socioeconomic status as defined by income, education, or occupation is associated with increased risk for oesophageal squamous cell carcinoma and, adenocarcinoma to a smaller extent.
7. ***Helicobacter pylori* infection:** Infection with *Helicobacter pylori*, and particularly with cagA+ strains, is inversely associated with the risk of adenocarcinoma of the oesophagus. Mechanism is not clear. In fact, it should not have been a factor responsible because *H. pylori* infection can cause chronic atrophic gastritis leading to decreased acid production, which negates the effects of chronic reflux, including the potential for development of Barrett's oesophagus.

SITES

- 50%—middle 1/3rd of oesophagus
- 33%—lower 1/3rd of oesophagus
- 17%—upper 1/3rd of oesophagus.

TYPES

I. Squamous cell carcinoma

1. Epitheliomatous ulcer (carcinomatous) with raised edges and flat base.
2. Proliferative growth (cauliflower) with surface ulcer which commonly bleeds.
3. Infiltrative variety or annular stenosing variety with a 5 years survival of 10%, gives rise to early dysphagia due to circumferential and longitudinal spread.
4. Polypoidal lesions (5 years survival 70%).

II. Adenocarcinoma

Mostly lower end and middle oesophagus (in Barrett's oesophagus).

CLINICAL FEATURES

- Men more than 60 years
- **Dysphagia** which is **progressive** and mainly for solids (it takes 18 months for dysphagia to develop). It means 60% of the circumference of the lumen is involved by the growth. By the time, the patient presents with dysphagia, the disease is fairly advanced. Hence, it has a poor prognosis. **Dysphagia is a late symptom because smooth muscle of oesophageal wall can dilate with ease due to lack of serosal layer on the oesophagus.**
- **Regurgitation** of the food contents. Haematemesis is not very common. Vomitus may contain streaks of blood and melaena is rare.
- Loss of appetite, loss of weight and cachexia
- Backache indicates enlarged lymph nodes (coeliac).

SPREAD

1. Local spread or direct spread

- To start with, it is a mucosal ulceration which spreads to the submucosa. Later, it causes fibrosis and the lumen gets narrowed. The spread occurs transversely and longitudinally. Once it spreads to all the layers, the structures in the vicinity are involved (Key Box 22.11).
- When the trachea is involved, tracheo-oesophageal fistula develops from carcinoma upper 1/3rd of oesophagus.
- Broncho-oesophageal fistula from carcinoma middle 1/3rd.
- Oesophagoaortic fistula results in massive bleeding (one of the causes of death).
- All these complications are contraindications for surgery and radiotherapy.

KEY BOX 22.11

FACTORS RESPONSIBLE FOR EARLY SPREAD AND AGGRESSIVE BEHAVIOUR OF CARCINOMA OESOPHAGUS

- Lack of serosal layer
- Proximity of vital structures
- Extensive mediastinal lymphatic drainage
- Late presentation

2. Lymphatic spread

Studies have showed that **patients having metastasis to 5 or fewer lymph nodes** have a better outcome.

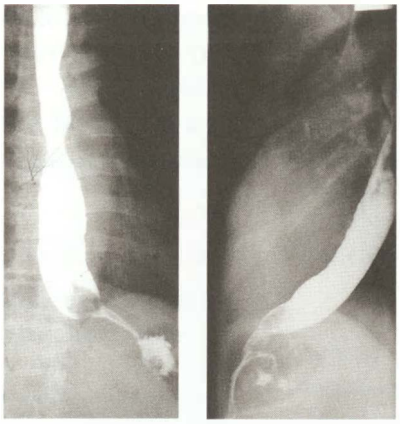
3. Blood spread

It results in secondaries in the liver, which clinically appear as nodular enlarged liver. Later, ascites and rectovesical deposits occur. Palpable left supraclavicular nodes indicate advanced disease. This sign is described as **Troisier's sign.**

INVESTIGATIONS

1. **Hb%** is low, which is the cause of generalised weakness.
2. **Liver function test (LFT)** is affected, if secondaries in liver occur (increased ALP).
3. **Ultrasound** is done to rule out liver secondaries, lymph nodes in the porta hepatis, coeliac nodes, etc.
4. **Barium swallow** demonstrates irregular, persistent, intrinsic filling defect (Figs 22.24 to 22.27)—shouldering is characteristic. Barium can also detect abnormal 'axis'. It can also demonstrate fistula. Both these features suggest that 'CURE' is not possible.
5. **Oesophagoscopy** to visualise the growth and to take biopsy (Figs 22.28 to 22.33 and Key Box 22.12).

Fig. 22.24: Barium swallow—carcinoma middle oesophagus



Figs 22.25 and 22.26: Carcinoma lower end of the oesophagus. Observe proximal shouldering

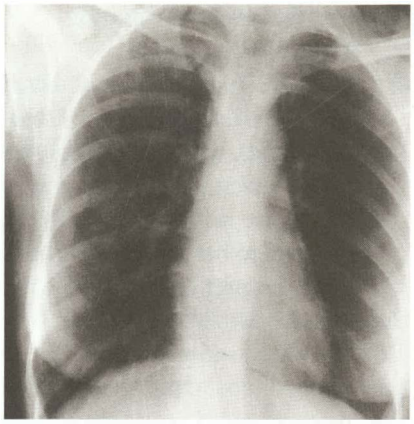


Fig. 22.27: Barium swallow showing straight axis—inoperable sign

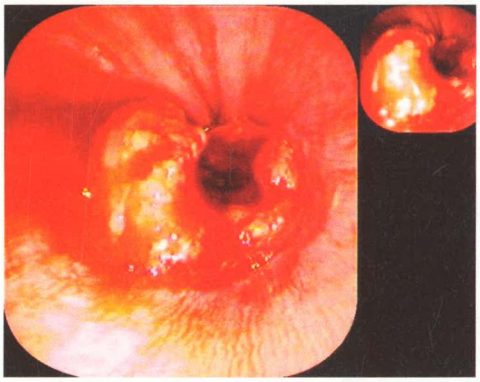


Fig. 22.28: Biopsy taken—observe the bleeding—friable growth

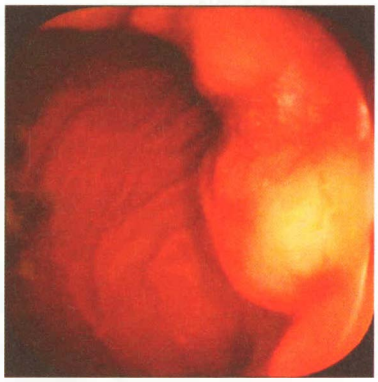


Fig. 22.29: Polypoidal growth at GE junction—patient presented with reflux

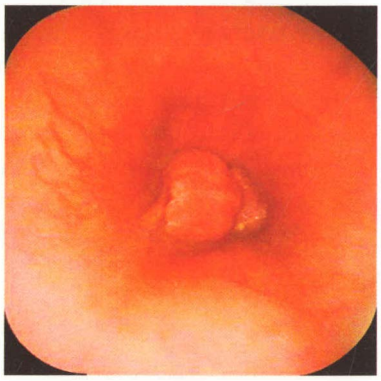


Fig. 22.30: Polypoidal growth obstructing the lumen of oesophagus

Fig. 22.31: Growth was extensive. Treated by total oesophagectomy

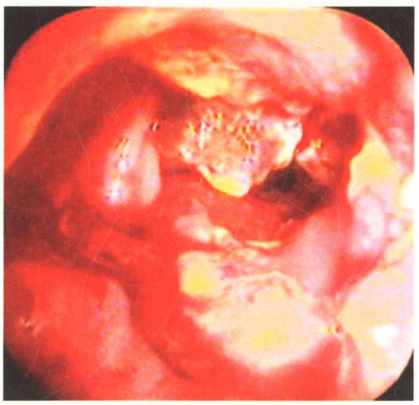


Fig. 22.32: Advanced carcinoma of GE junction

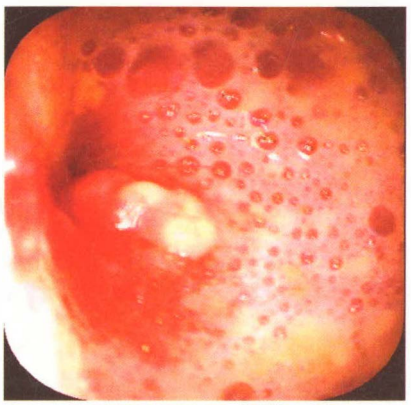


Fig. 22.33: Ca-midoesophagus—obstruction with stasis of food and saliva

- **Multiple biopsies** may be necessary. In high-risk areas like China, **endoscopic staining with supravital dyes** (indigocarmine) is done to identify dysplastic epithelium. If found, some advocate endoscopic mucosal resection (Key Box 22.12).

PEARLS OF WISDOM

Dysplasia in Barrett's mucosa is a prognostic sign of impending malignant degeneration.

6. **Chest X-ray** to rule out aspiration pneumonia, mediastinal widening and posterior tracheal indentation.
7. **Bronchoscopy** to rule out involvement of bronchus, as in carcinoma middle 1/3rd. If involved, it can confirm that tumour is locally unresectable.
8. **CT scan** (Figs 22.34 and 22.35) of the chest to detect local infiltration. It is a very useful investigation before contemplating for total oesophagectomy. It is useful to

KEY BOX 22.12

CHROMOENDOSCOPY

- Chromoendoscopy involves the topical application of stains or pigments to improve tissue localisation, characterisation, or diagnosis during endoscopy.
- Several agents have been described that can broadly be categorised as absorptive (vital) stains, contrast stains, and reactive stains.
- Absorptive stains (e.g. Lugol's solution and methylene blue) diffuse or are preferentially absorbed across specific epithelial cell membranes.
- Contrast stains (e.g. indigocarmine) highlight surface topography and mucosal irregularities by permeating mucosal crevices.
- Reactive stains (e.g. Congo red and phenol red) undergo chemical reactions with specific cellular constituents, resulting in a colour change.

assess involvement of vital structures such as bronchus, airway, pericardium, aorta, etc. Obliteration of fat planes gives the clue

9. **Endoscopic ultrasound** to know the depth of wall involvement, to detect mediastinal lymph nodes, etc (Fig. 22.36 and Key Box 22.13).

KEY BOX 22.13

ENDOSCOPIC ULTRASOUND (ENDOSONOGRAPHY)

- It relies on a high frequency (5–30 MHz) transducer
- To determine the depth of spread of a malignant tumour through the oesophageal wall
- Involvement of adjacent organs
- Metastasis to lymph nodes
- Also detects contiguous spread ***downward into cardia***
- Can detect metastasis in the liver
- Can also detect small lymph nodes which are less than 5 mm (which cannot be detected by CT scan also)
- If 'endosono' detects more ***than 5 lymph nodes,*** it is not a case for curative resection.

10. **Positron emission tomography (PET)**/computerised tomography scanning: PET scan is based on the principle that metabolically active tumour cells can be visible once radiopharmaceutical agent 18F-fluorodeoxyglucose (FDG) is given intravenously. These lymph nodes, if malignant are clearly visible thus differentiating them from nonmalignant enlargement.

    Applying the same principles, PET can be used to know the 'response' after radiotherapy or chemotherapy.

11. **Thoracoscopy and laparoscopy** At the end of all investigations, it is important to decide one can resect this or not? Is it worthwhile resecting? Can patient tolerate the procedure? Even in the absence of systemic spread, some features will suggest surgical cure is unlikely (Key Boxes 22.14 to 22.16).

KEY BOX 22.14

UNLIKELY SURGICAL CURE IN THE ABSENCE OF SYSTEMIC SPREAD

- **L**oss of weight more than 20%
- (O)**E**sophageal 'axis' is abnormal on barium test
- **N**odes—multiple on CT scan
- **G**rade—invasive, poorly differentiated
- **T**umour length more than 8 cm
- **H**orner's syndrome

Remember as **LENGTH**

KEY BOX 22.15

THORACOSCOPY AND LAPAROSCOPY

- Staging tools in oesophageal cancer.
- More accurate in determining nodal status than non-invasive techniques and helpful in evaluating the extent of local invasion and metastatic disease.
- Thoracoscopy is accurate in 93% and laparoscopy in 94% of patients in identifying metastatic disease.
- 6 cases of unsuspected coeliac nodal disease were identified in 19 patients despite preoperative CT and EUS.

KEY BOX 22.16

LAPAROSCOPIC ULTRASOUND (LUS)

- LUS is also a staging modality and may provide improved accuracy in T and N staging.
- LUS may also be more accurate for staging coeliac nodes because EUS probe is more distant and does not provide direct inspection.

Depth of invasion

- For T1 lesions, conservative oesophageal resection such as vagal-sparing, transhiatal or minimally invasive oesophagectomy can be opted.
- For localised intramucosal tumours of limited extent, EMR or ESD are an acceptable alternative.
- There is almost no role for chemoradiotherapy for treatment of T1 lesions.
- Surgical or endoscopic resection alone carries a good long-term survival, as high as 88% in some series.

DEPTH OF TUMOUR vs LYMPH NODE INVOLVEMENT

1.	T1 lesion (intramucosal)	18% LNI
2.	T1 lesion (submucosal)	55% LNI
3.	T2 lesion	60% LNI
4.	T3 lesion	80% LNI

General assessment for staging

- General health of the patient, co-morbid conditions, fitness for anaesthesia—all these factors decide the choice of therapy.
- TNM staging
- In the absence of systemic spread, surgical cure may not be possible as depicted in Key Box 22.14.

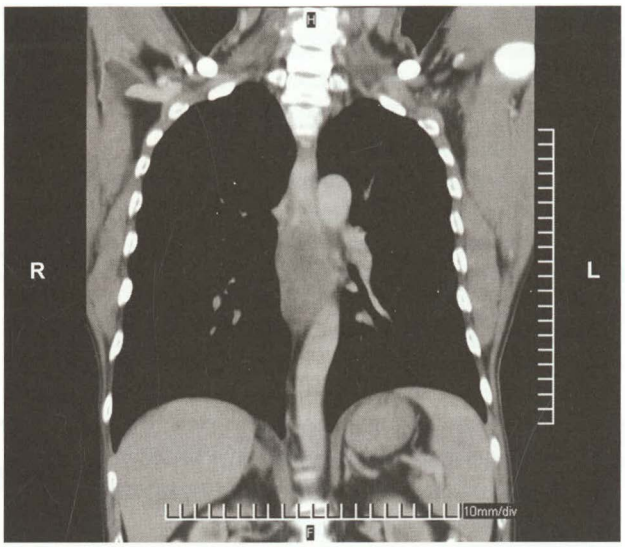


Fig. 22.34: Contrast CT showing a large midoesophageal growth

Fig. 22.35: Contrast CT lateral picture showing total occlusion of midoesophagus

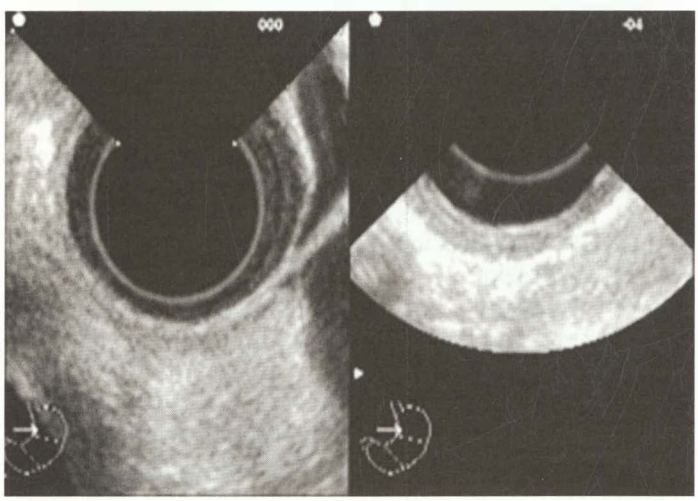


Fig. 22.36: Endosonography showing oesophagus and stomach layers (*Courtesy:* Prof Ganesh Pai, HOD of Medical Gastroenterology, KMC, Manipal)

TNM STAGING AJCC 7th edition

Tumour

T0 No primary tumour
Tis Carcinoma *in situ*—high-grade dysplasia
T1a Tumour invades lamina propria, muscularis, mucosa
T1b Tumour involving submucosa
T2 Tumour involving muscularis propria
T3 Tumour with perioesophageal spread
T4 Involvement of recurrent laryngeal nerve, phrenic nerve, sympathetic chain, azygos vein; malignant effusion (adjacent structures)

Nodal status

Nx Lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Regional lymph node metastasis—1 to 2 nodes
N2 Metastasis in 3–6 regional nodes
N3 Metastasis in 7 or more nodes

Metastasis

M0 No distal spread
M1 Upper thoracic oesophageal carcinoma with spread to other nonregional nodes or distant spread. Middle thoracic oesophageal carcinoma with spread to neck nodes/ coeliac nodes or other nonregional nodes. Lower thoracic oesophageal carcinoma with spread to other nonregional nodes or distant spread.
Mx Distant metastasis cannot be assessed.

TNM Stage Grouping

Stage 0 – Tis N0 M0
Stage 1 – T1 N0 M0
Stage IIA – T2 N0 M0, T3 N0H0
Stage IIB – T1N1N0, T2N1, M0
Stage III – T3N1M0, T4 and N1N0
Stage IV – A–Any T and N, M1a
B–Any T1 and N, M1b

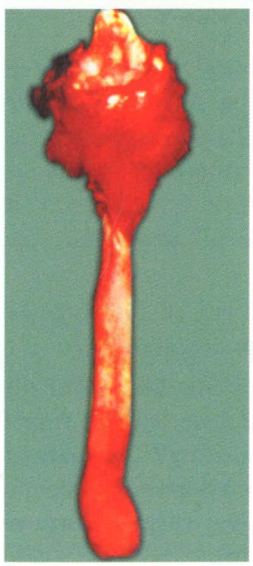


Fig. 22.37: Total pharyngolaryngo-oesophagectomy—followed by gastric pull-up

TREATMENT

I. A few considerations before treatment of carcinoma—since they often present at an advanced stage, 5-year survival is very low in the majority of cases. The following factors are considered for *palliative treatment:*

1. Comorbid conditions:	Cardiac diseases, bad chest
2. Blood spread metastasis:	Para-aortic lymph nodes
3. Contiguous organ invasion (CT scan):	Trachea, aorta, pericardium
4. Diagnostic laparoscopy:	Peritoneal spread Omental nodules ascites
5. CT scan or laparoscopic ultrasound	Lymph nodes metastasis: Para-aortic lymph nodes supraclavicular lymph nodes

II. Treatment with curative intent

- Squamous cell carcinoma is the most common type identified in patient with achalasia cardia.

1. Carcinoma cervical and upper thoracic oesophagus

- In vast majority of cases, treatment of carcinoma oesophagus is aimed at to relieve dysphagia by palliative resection/palliative radiotherapy/metallic stenting.
- **Tumours that have not penetrated** through oesophageal wall and have not metastasised to regional lymph nodes are **potentially curable.**
- **Decision of surgery/radiotherapy** also depends upon location of the tumour, histology, site of the tumour, staging and cardiopulmonary reserve. Nutritional status has to be improved to raise albumin levels from 3 to 3.4 g/dl to decrease complications such as anastomotic leak.
- **Cervical oesophageal cancers** (Fig. 22.37) are almost always squamous cell carcinoma. Usually they are postcricoid in a female patient. It is very unusual for this malignancy to involve intrathoracic nodes. Upper thoracic is also squamous cell carcinoma. Involvement of thoracic nodes, early spread to trachea, bronchus and great vessels is common.
- Surgery followed by radiotherapy is the treatment of choice. Local recurrence is high and they succumb to local disease.
- If adjacent structure is not involved, total oesophagectomy with or without laryngectomy can be done. Preoperative neoadjuvant chemotherapy can also be given first (2–3 cycles followed by 3–5 Gy of radiation therapy).
- This is followed by total laryngectomy with bilateral neck dissection and reconstruction using gastric pull-up.
- Tumours in the upper thoracic oesophagus are removed by posterolateral thoracotomy, excision of the tumour and lymph nodes followed by gastric pull-up requiring laparotomy and pharyngogastric anastomosis in the neck. This is the three-stage oesophagectomy.

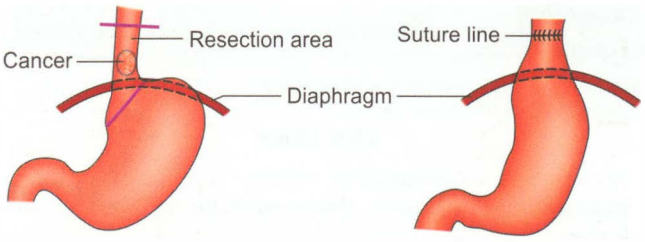


Fig. 22.38: Ivor-Lewis operation

Fig. 22.39: Anastomosis in the thorax

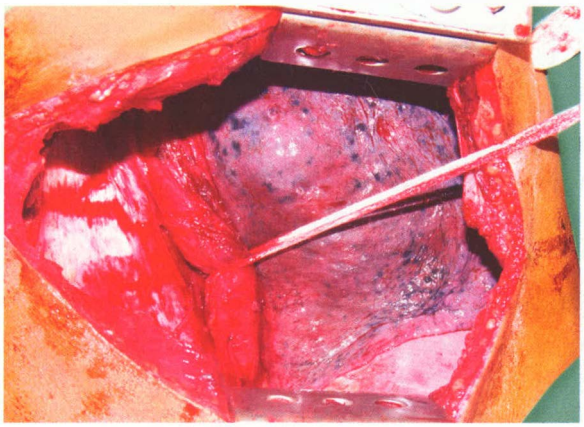


Fig. 22.40: Oesophagus is mobilised in the thorax

2. Tumours of the thoracic oesophagus and cardia

They are usually **adenocarcinomas**. These tumours have a tendency to spread for long distances submucosally and the lymphatic flow is in the longitudinal direction.

Two choices (Table 22.6)

A. Ivor-Lewis operation

- In this operation, abdomen is opened first, stomach is mobilised and the wound is closed. The patient is put in left lateral position, and right thoracotomy is done through 6th intercostal space. The growth is removed and oesophagogastric anastomosis is done inside the thorax, above the level of aortic arch. Hence, it is described as a two-stage Ivor-Lewis approach (Figs 22.38 to 22.40).
- However, a few surgeons do not advocate this surgery because of following reasons:
  1. Higher incidence of **pulmonary complications**.
  2. Higher morbidity associated with **anastomotic leak**.
  3. Higher incidence of **oesophagitis**.
  4. Higher chances of **local recurrence** due to inability to resect long length of oesophagus.

  Hence, the alternate procedure (McKeown) is a better choice.

B. McKeown's three stage *en bloc* oesophagectomy

En bloc resection is the best treatment with removal of all lymph nodes (abdominal and thoracic). Hence, three incisions are required (1) abdominal, (2) right posterolateral thoracotomy through the fifth space and (3) left cervical incision. Local recurrence at the anastomosis is not a

Table 22.6 Total oesophagectomy—comparison of two types

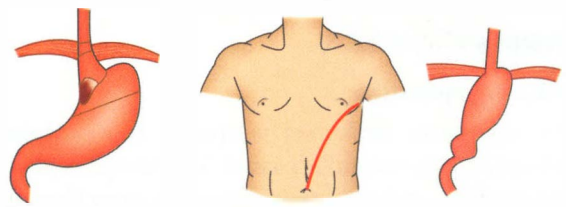
En bloc oesophagectomy—McKeown	Transhiatal resection—Orringer
First choice	**Alternate choice**
• Three incisions: Consequently, mobility is restricted and postoperative pain is more. Hence, morbidity and mortality are higher	• Two incisions: Abdominal and cervical. Hence, mobility is better and postoperative pain is less
• Under vision all lymph nodes in the thorax, neck and the abdomen can be removed	• Pulmonary complications are less as chest is not opened. Hence, mortality and morbidity is less than *en bloc* resection
• Surgery takes longer period of time	• Some mediastinal lymph nodes get cleared if proper dissection is done through hiatus
• However, long-term survival is not much compared to THE	• Surgery takes much shorter period of time
• When lymph nodes involved are less than 8 in number, survival is better than THE	

problem because of wide clearance (by obtaining a 10 cm margin of normal oesophagus above the tumour). Gastrointestinal continuity is obtained by **gastric tube anastomosis to cervical oesophagus.**

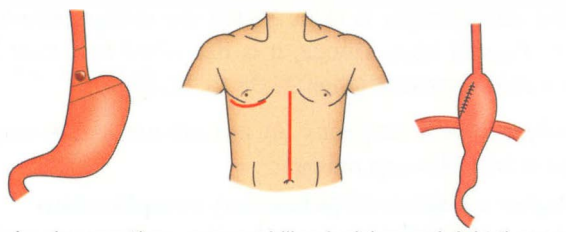
3. Carcinoma lower oesophagus and cardia

- They are adenocarcinomas.
- Surgery is the best method of treatment and palliation.
- *En bloc* resection is the best treatment with removal of all lymph nodes (abdominal and thoracic).
- An alternative method is **transhiatal oesophagectomy** (THE) without thoracotomy.

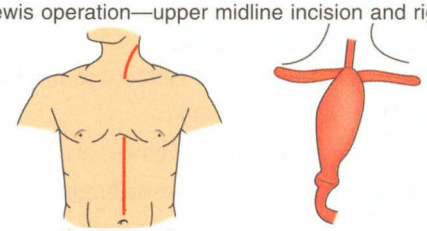
Figure 22.41 shows various types of oesophageal surgery done for carcinoma oesophagus.



Left thoracoabdominal incision for carcinoma GE junction growth



Ivor-Lewis operation—upper midline incision and right thoracotomy



Orringer's transhiatal oesophagectomy—upper midline incision and incision in the left side of the neck along the sternomastoid muscle and anastomosis at cervical oesophagus to stomach.

Fig. 22.41: Various incisions for oesophageal surgery

PALLIATIVE PROCEDURES

1. Intubation: The choice of the intubation material is self expanding metal stent (SEM). Two types are available—covered and noncovered. The stent is deployed by endoscopy under fluoroscopic guidance. It is collapsed during insertion and once confirmed radiologically by position, it is expanded. Minimal dilatation of the oesophagus (8 mm) is required to place the stent (Figs 22.48 to 22.51). On the other hand, the plastic tubes which were used earlier, required more than 8 mm dilatation and there was always a risk of perforation. If the patient is expected to survive beyond 3 months, SEM stent can be considered. Complications include aspiration, feeling of tightness in the chest, bleeding, food blockage, etc.

- This is the choice in cases of tracheo-oesophageal fistula due to carcinoma oesophagus.

2. Laser therapy: Endoscopic laser treatment is used to core the tumour and widen the lumen to relieve dysphagia. Principle is thermal destruction of the tumour. The procedure has to be repeated. Laser used is Nd:YAG laser or diode laser. Infiltrative lesions are not suitable for laser treatment but exophytic lesions are ideal—less than 6 cm.

- Success rates are 80–90% in relieving dysphagia.
- Perforation, fever, chest pain, fistula are complications.

3. Photodynamic therapy: It is used in early carcinomas—mucosal lesions wherein patients are not willing for surgical procedures including endoscopic mucosal resection. In this procedure, a photosensitiser (porfimer sodium) is given. It will be taken by dysplastic cells and malignant cells.

- Perforation, fever, sunburn, pleural effusion are the complications.

4. Radiotherapy, brachytherapy: It is intraluminal radiation (RT) with short penetration distance 1500 cGy radiation is given. No systemic side-effects. Palliative external beam radiation to doses of 50 to 60 Gy is successful in 50 to 70% of patients.

5. Intratumour injection of absolute alcohol under endoscopic control: Used as an adjunct to stenting to deal with tumour overgrowth.

TRANSHIATAL OESOPHAGECTOMY (THE)—WITHOUT THORACOTOMY—ORRINGER (Figs 22.42 to 22.47)

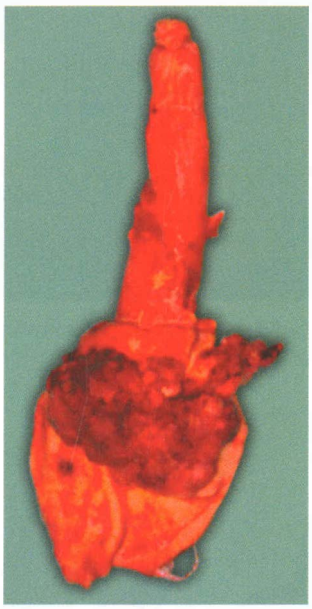


Fig. 22.42: Transhiatal oesophagectomy (THE) specimen done for Ca GE junction

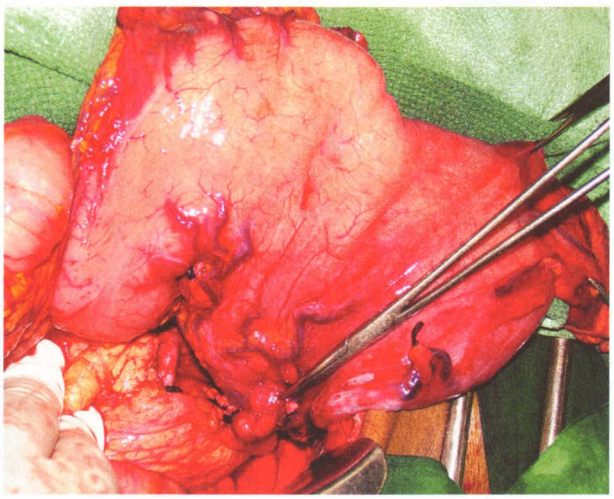


Fig. 22.43: Stomach mobilised by ligating left gastric pedicle

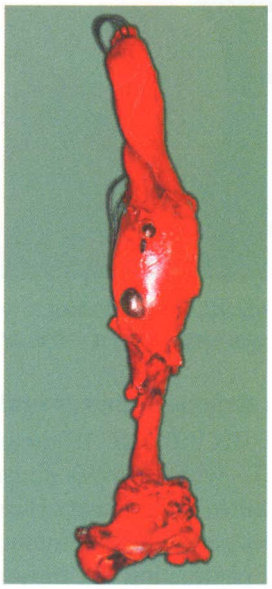
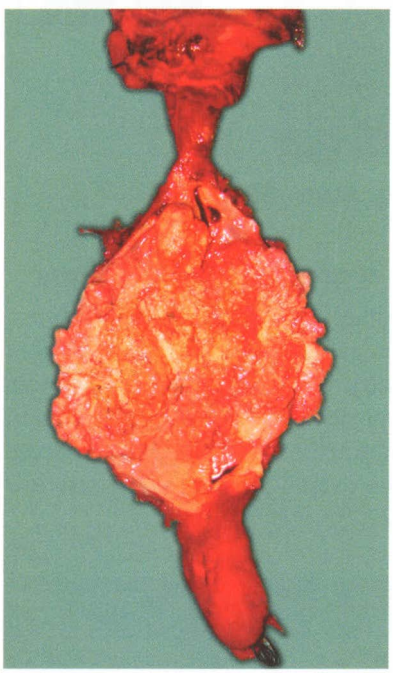
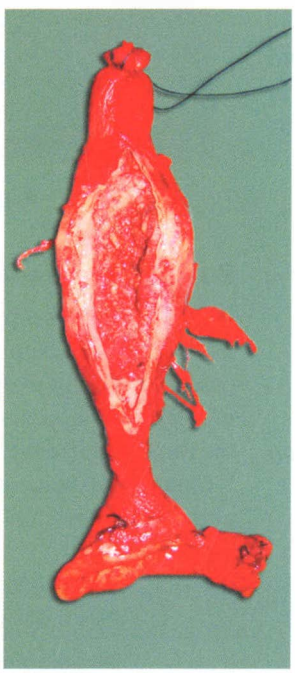


Fig. 22.44: THE for mid-oesophageal cancer





Figs 22.45 and 22.46: Opened specimen showing extensive ulceration and nodularity—minimum 10 cm clearance for oesophageal cancer is important. Hence, THE is better than Ivor-Lewis operation to get this length (*Courtesy:* Dr Basavananda Hartimath, Associate Professor, Department of Surgery, KMC, Manipal)

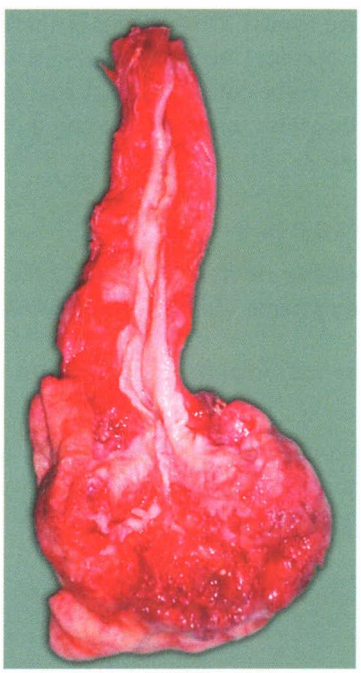


Fig. 22.47: Transhiatal (Orringer) total oesophagectomy (*Courtesy:* Dr Sachidanand, Associate Professor, Karnataka Institute of Medical Sciences, Hubli, Karnataka)

Carcinoma lower end of oesophagus—without opening the thorax by mobilisation from above and below, the oesophagus with the growth can be removed.

Comments: Transhiatal oesophagectomy is a very good alternative (almost as good as *en bloc*) to lower oesophageal and GE junction tumours. No specific attempt is made to remove lymph nodes. Patients tolerate the procedure very well. No mediastinal complications related to leak are seen because of anastomosis in the neck. It gives a reasonably good relief from dysphagia and survival. It is not blind oesophagectomy. With good retraction of the hiatus, mobilisation can be done under vision. It is not the type of operation that influences survival but rather the stage of the disease at the time of the operation is performed.

- Prophylactic vagal sparing THE is indicated in severe dysplasia and intramucosal lesions.

Fig. 22.48: Endoscopic view of carcinoma lower oesophagus—it is an adenocarcinoma

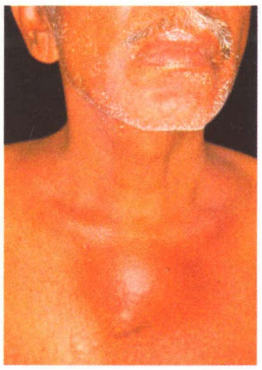


Fig. 22.49: Carcinoma oesophagus with the sternal secondary

Fig. 22.50: Oesophageal stenting with metallic stent

6. Argon plasma coagulation

7. BICAP: BICAP tumour probes (Circon-ACMI, Stamford, CT), which provide direct application of electrical current, are limited to treatment of tumours that are circumferential. Bipolar electrocoagulation tumour probe was employed for palliation of obstructing circumferential oesophageal cancer.

8. Surgical palliation

- **Resection:** In fact transhiatal oesophagectomy is considered as palliative treatment
- **Bypass:** Palliative bypass: Oesophagus is not resected—gastric tube to cervical oesophagus—mortality and morbidity is high. To be considered last, that too only in expert hands. Otherwise, consider feeding jejunostomy.

9. Feeding gastrostomy or jejunostomy

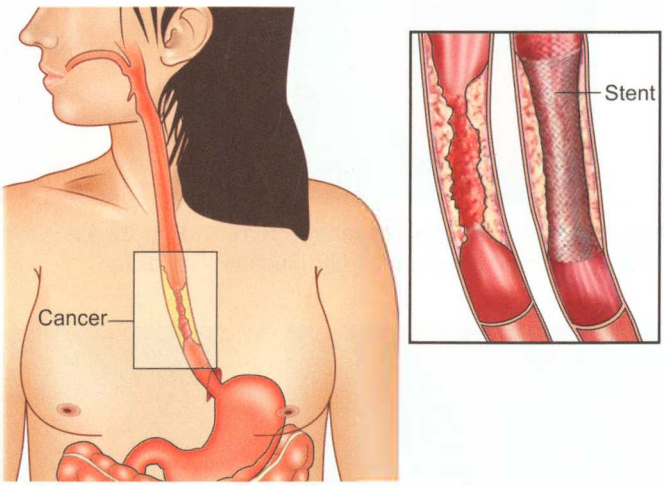


Fig. 22.51: Stent—SEMS

PEARLS OF WISDOM

Metallic self-expandable stents are currently the choice of tubes to relieve dysphagia and to achieve palliation.

Causes of death in carcinoma oesophagus

- Cancer cachexia
- Complications such as bronchopleural fistula, aspiration pneumonia, haematemesis due to erosion of aorta, perforation of the growth and mediastinitis.

Summary of treatment of carcinoma oesophagus

In spite of endoscopy and other investigations, the treatment of carcinoma oesophagus is palliative in more than 95% of cases, especially in our country. By the time the first symptom of dysphagia appears, it is too advanced. Also, the location of the tumour, adjacent structures in the mediastinum, widespread lymphatic drainage alters the overall prognosis to a very low percent of survival (Table 22.7).

Table 22.7 Comparison of two types of oesophageal cancer

	Squamous cell carcinoma	Adenocarcinoma
Site	Upper two-thirds	Lower one-third
Predisposing factors	Smoking alcohol	Barrett's oesophagus—GORD
Behaviour	Less aggressive	More aggressive
Incidence	Less common	More common
Achalasia cardia	Is the predisposing factor	Does not give rise to adenocarcinoma
Chemoradiation	It can be an alternate treatment	Not alternate to surgery
Surgery	Can be the first choice or chemoradiation	Surgery is the first choice—unfit patient, chemotherapy
Dietary carcinogens	A. Elevated nitrates in drinking water B. Food containing fungi-*Geotrichum candidum*—pickles, corn C. Baked bread	Consumption of meat Alcohol

CERTAIN FACTS

- Majority of oesophageal cancers are advanced at the time of diagnosis.
- 5 years survival rates after curative resections are around 10%.
- **The best results are obtained after surgery—radial oesophagectomy.**
- Radiotherapy, chemotherapy, dilatation and stenting also can give palliation.
- Curative resections are major surgeries and should be undertaken by an experienced surgeon.
- Curative resections can be attempted at all levels of carcinoma oesophagus (upper, middle and lower) provided vital structures are **not involved** (assessment by CT scan and endosonogram).
- **Chemoradiotherapy may cure the disease** in selected patients—these squamous cell cancers.
- **Palliation:** As majority of the patients with carcinoma oesophagus have dysphagia, all the methods of palliation are aimed at relieving dysphagia. Endoscopy followed by dilatation using guide wire is a simple procedure with rate of complications such as perforation around 2–3%. However, the results are only for a few weeks. Self-expandable metal stents are regularly used nowadays as a palliation. They have two layers of superalloy monofilament wire with a layer of silicon between them. This prevents the tumour from growing within the stent.
- **Endoscopic laser therapy** relieves obstruction and bleeding. It can be carried out as an outpatient treatment but needs multiple sittings.
- **Radiotherapy** alone has been tried for squamous cell carcinoma of the oesophagus—dose is 6,000 cG units. No survival advantage is seen with this method.
- **Combination chemotherapy** using cisplatin and 5-fluorouracil with radiotherapy has been tried in patients who are not fit for surgery. Paclitaxel achieves high response rate in metastatic oesophageal cancer and it also acts as a radiation sensitiser.
- **Endoscopic intraluminal brachytherapy** is used in cases of recurrent tumour growing inside the lumen and causing obstruction.

PEARLS OF WISDOM

The entire treatment of carcinoma of oesophagus is aimed at 'Cure' in minority of cases and relief of dysphagia in almost all cases.

OESOPHAGEAL STRICTURE (Key Box 22.17)

CORROSIVE INJURY

These caustics are taken accidentally or in attempted suicide. The agents are sodium hydroxide, sulphuric acid, household bleach.

KEY BOX 22.17

CAUSES OF STRICTURE

1. Corrosive (caustic) injuries
2. Carcinoma
3. Columnar lined oesophagus—Barrett's
4. Capsules of tetracycline group of antibiotics
5. C vitamins—potassium compounds
6. Chronic reflux due to GORD, results in **Schatzki ring**—a stricture at lower oesophagus at squamocolumnar junction. It is a circular ring consisting of fibrous tissue and cellular infiltrates.

- The acids affect acidic stomach mucosa.
- The squamous epithelium of oesophagus is relatively resistant to acids (also due to rapidity of flow of fluids).
- The alkali—sodium hydroxide affects oesophageal mucosa.
- The corrosive injuries may involve oropharynx, larynx, oesophagus, stomach and sometimes intestines also.
- Alkali ingestion is more common than acid ingestion
- Alkali ingestion is more devastating than acid ingestion
- Alkali ingestion almost always leads to significant destruction of oesophagus
- Alkali affects alkaline mucosa, oesophagus more than stomach mucosa.

Classification of injuries

- **Superficial:** Erythema, oedema, blisters, etc. Re-epithelialisation of mucosa is complete by 6th week.
- **Deep:** Circumferential ulcers, produces scarring and contractures. Major injury of the stomach produces gastric outlet obstruction.

Clinical features

- Severe pain, drooling of saliva, inability to swallow.
- Retrosternal burning, abdominal guarding and rigidity.
- Hoarseness, stridor, laryngeal oedema, if there is laryngeal injury.
- Dysphagia occurs later due to stricture.

PEARLS OF WISDOM

- ***What should not be done: Stomach wash or induced vomiting, as it will aggravate oesophageal injury or it may result in aspiration. No role for corticosteroids in cases of deep injuries or acid injuries.***
- ***What should be done: Verification of the aetiologic agent and accurate assessment of depth and extent of injury.***

Important examinations in corrosive injury

1. Examination of the oral cavity, chest and abdomen.
2. Auscultation of the lungs
3. Examination of upper airway

4. Abdominal examination—guarding and rigidity of the abdomen indicate perforation.

Investigation

1. Early endoscopy—within 12–24 hours—to grade the injury (*see* below for detail).
2. CT scan can reveal any perforation in the thorax or in the abdomen.

Treatment

Can be divided into acute and chronic cases. In acute cases, treatment depends upon the severity of the injury or burn

Acute cases: Any surgical intervention should be done only in selected referral centres. Kindly follow the instructions given below.

TEN COMMANDMENTS OF ACUTE CORROSIVE INJURY

1. Neutralisation of the ingested material
2. Alkalis are neutralised by half strength vinegar, citrus juices
3. Acids are neutralised by milk, egg white, antacids
4. Avoid emetics and sodium bicarbonates
5. Stabilise the patient and arrange upper GI scopy—classify the degree of injury.
6. **No burns:** Observe, small amount of clear fluids may be allowed orally
7. **First degree burns:** Observation for 48 hours. If he can swallow saliva, clear fluids can be started. Repeat endoscopies after a month, 3 or 6 months later to rule out stricture.
8. **Second degree burns:** Intravenous fluids, nil per oral, acid suppression, relieve airway obstruction, parenteral nutrition
9. Cervical oesophagostomy, staple the GE junction, feeding jejunostomy. 6 weeks later, gastric pull up and oesophagogastric anastomosis, in cases of perforation.
10. Call for senior surgeon's help

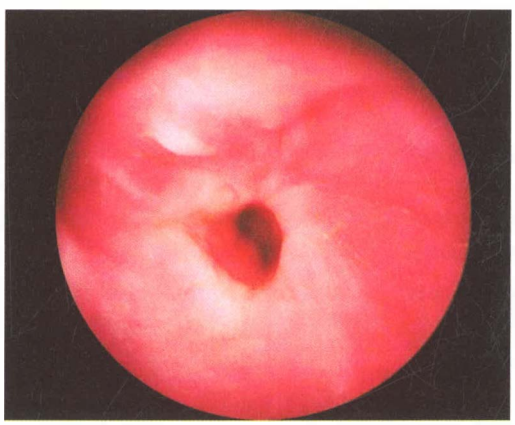


Fig. 22.52: Endoscopic view of corrosive strictures of lower end of oesophagus. The scope could not be negotiated through the oesophagus. He underwent dilatation. At least 16 mm diameter of the oesophagus is the requirement for normal swallowing

Chronic cases present with **stricture** that need regula dilatation. This is ideally done after 6 weeks (the time for re epithelialisation of oesophagus) (Figs 22.52 to 22.54 an Key Box 22.18).

- **Colonic pull up** is the choice for impassable strictures, onl after adequate nutrition is achieved through feedin jejunostomy.

KEY BOX 22.18

DILATATION OF THE STRICTURE

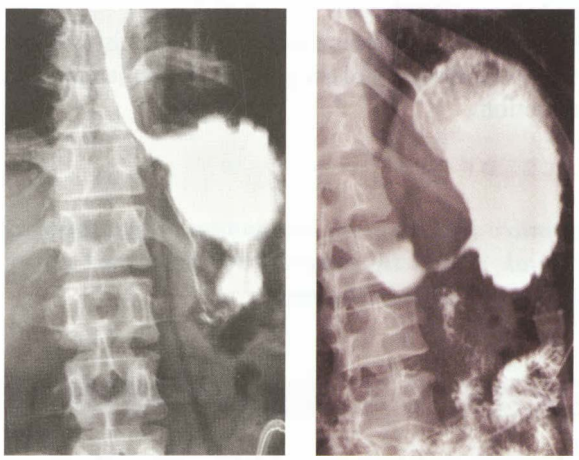
- Indicated in all cases of symptomatic oesophageal stricture.
- Peptic strictures, corrosive strictures, anastomotic strictures following oesophagogastric anastomosis in the neck after 'THE' or following oesophagogastrectomy for lower oesophageal cancer.
- Flexible oesophagoscopy is done and a guide wire is passed. The scope is withdrawn. Solid dilators of increasing diameter are passed over the guidewire.
- A stricture should be dilated to **at least 16 mm** in diameter to restore **normal swallowing**.
- Peptic strictures are **easy to dilate** as they are **short**.
- Beware of a GE junction stricture. It may be malignant. Once it is dilated, a growth may be seen. Take a biopsy.
- Pneumatic dilatation can also be used.

Complications of stricture

- **Development of malignancy** (3 to 5%)
- Progressive nutritional deficiency
- Recurrent respiratory tract infection.

OESOPHAGEAL PERFORATIONS

- Oesophageal perforation is a serious, acute emergency carrying a very high mortality rate. Fortunately it is not common. There are many causes and are totally different from gastric or duodenal perforations.



Figs 22.53 and 22.54: Corrosive strictures of lower end of oesophagus and pyloric antrum of stomach. Oesophageal stricture was dilated. However, antral stricture could not be dilated. Hence, he underwent vagotomy and gastrojejunostomy

- Instrumentation perforations are the most common cause of perforation.
- **Cervical oesophagus** is the most likely site of perforation.
- **Cricopharyngeal** region is the most common site.

Causes

1. **Instrumentation:** Endoscopy, dilatation of strictures, injection sclerotherapy and laser treatment are the common causes.
2. **Operative:** Thyroidectomy, vagotomy, spine surgery, mediastinoscopy.
3. **Traumatic:** Caustic injury, sharp and blunt trauma
4. **Oesophageal diseases:** Carcinoma, Barrett's oesophagus
5. **Spontaneous rupture:** Boerhaave's syndrome.

Clinical features

- **Severe pain in the neck,** chest or abdomen, stiffness of the neck depending upon the site of perforation.
- Haematemesis, dysphagia, dyspnoea, hypotension, **tachycardia, shock** and pleural effusion are the other features.
- **Mackler's triad:** Chest pain, vomiting and subcutaneous emphysema. It is present only in 10 to 15% of patients.
- **'Hamman's sign'** escape of air into the mediastinum resulting in "mediastinal crunch" which is produced by the heart beating against air-filled tissues.

Investigations

- The investigation of choice is plain chest X-ray which can demonstrate **pneumomediastinum,** subcutaneous emphysema and mediastinal air fluid levels. When in doubt, **CT chest** is diagnostic.
- **Contrast swallow** can also be done.

Treatment principles

- Infection is polymicrobial: Staphylococcus, Streptococcus, Pseudomonas and Bacteroides.
- **Early surgery** should be done within 24 hours. Closure of perforation and external drainage is the treatment.
- Perforation older than 24 hours—treated by temporary cervical oesophagostomy, ligation of oesophagus proximal to GE junction and feeding jejunostomy. After 6–8 weeks, oesophagectomy and gastric or colonic pull up is done.
- **Conservative treatment has risk of continuing leak, sepsis and mediastinitis.**

BAROTRAUMA – BOERHAAVE SYNDROME

Spontaneous perforation

It occurs when a person vomits against a closed glottis.

Historical: First reported by Professor Hermann Boerhaave—a case of grand admiral who was a glutton and practising autoemesis.

Aetiopathogenesis

- Rupture is due to sudden rise in oesophageal pressure during vomiting.
- Excessive eating as in bulimia/excessive alcohol has been thought to be responsible.
- It is a serious condition because of large volume of material realised under pressure.

Site of perforation

Posterolateral wall of the lower third of the oesophagus.

Investigations

- Chest X-ray Air in the mediastinum
  Air in the pleura
  Air in the peritoneum
  Pleural effusion
- CT scan It is the investigation of choice to detect the exact site of perforation

Treatment principles (as described earlier)

DIVERTICULUM OF OESOPHAGUS

Types of diverticulum (Table 22.8)

- **Cervical diverticulum** (Fig. 22.55) is the commonest type of diverticulum which can present with regurgitation of meals, aspiration of food contents into the lungs or recurrent respiratory tract infections. It is treated by excision of the sac and repair of the defect along with cricopharyngeal myotomy—**posterior midline**.

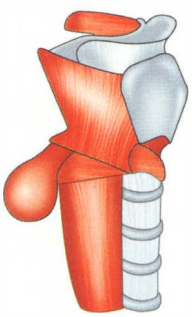


Fig. 22.55: Diverticulum

Table 22.8 Types of diverticulum

Name	Situation	Aetiology
1. **Zenker's** diverticulum (**cervical** or pulsion)	Proximal to the upper oesophageal sphincter	Protrusion of oesophageal mucosa between cricopharyngeus muscle inferiorly and thyropharyngeus muscle superiorly
2. Epinephric diverticulum	Proximal to the lower oesophageal sphincter	It is due to some kind of motility disturbance, wherein protrusion occurs in the lower end
3. Parabronchial diverticulum	Midoesophagus	Tuberculosis, by causing enlargement of mediastinal nodes, fibrosis and adhesions produce traction on the oesophagus, which results in the diverticulum

DIFFERENTIAL DIAGNOSIS OF DYSPHAGIA (Fig. 22.56)

I. Causes from outside (extraluminal) oesophagus

- Thyroid swellings
- Cardiomegaly
- Aortic aneurysm
- Mediastinal nodes: Tuberculosis, lymphoma or secondaries
- Rolling hiatus hernia

II. Causes in the wall of oesophagus (luminal)

1. Oesophageal stricture
   - Corrosive acid poisoning
   - Tuberculous stricture
2. Carcinoma oesophagus
3. Oesophageal diverticulum
4. Muscular spasm: Plummer-Vinson syndrome and achalasia cardia.
5. Tetanus

III. Causes in the lumen of oesophagus

1. Foreign body: Dentures, coins, etc.

History

1. Acute dysphagia: Common in children. Foreign bodies are common causes. Acute dysphagia with pain suggests tonsillitis or pharyngitis.

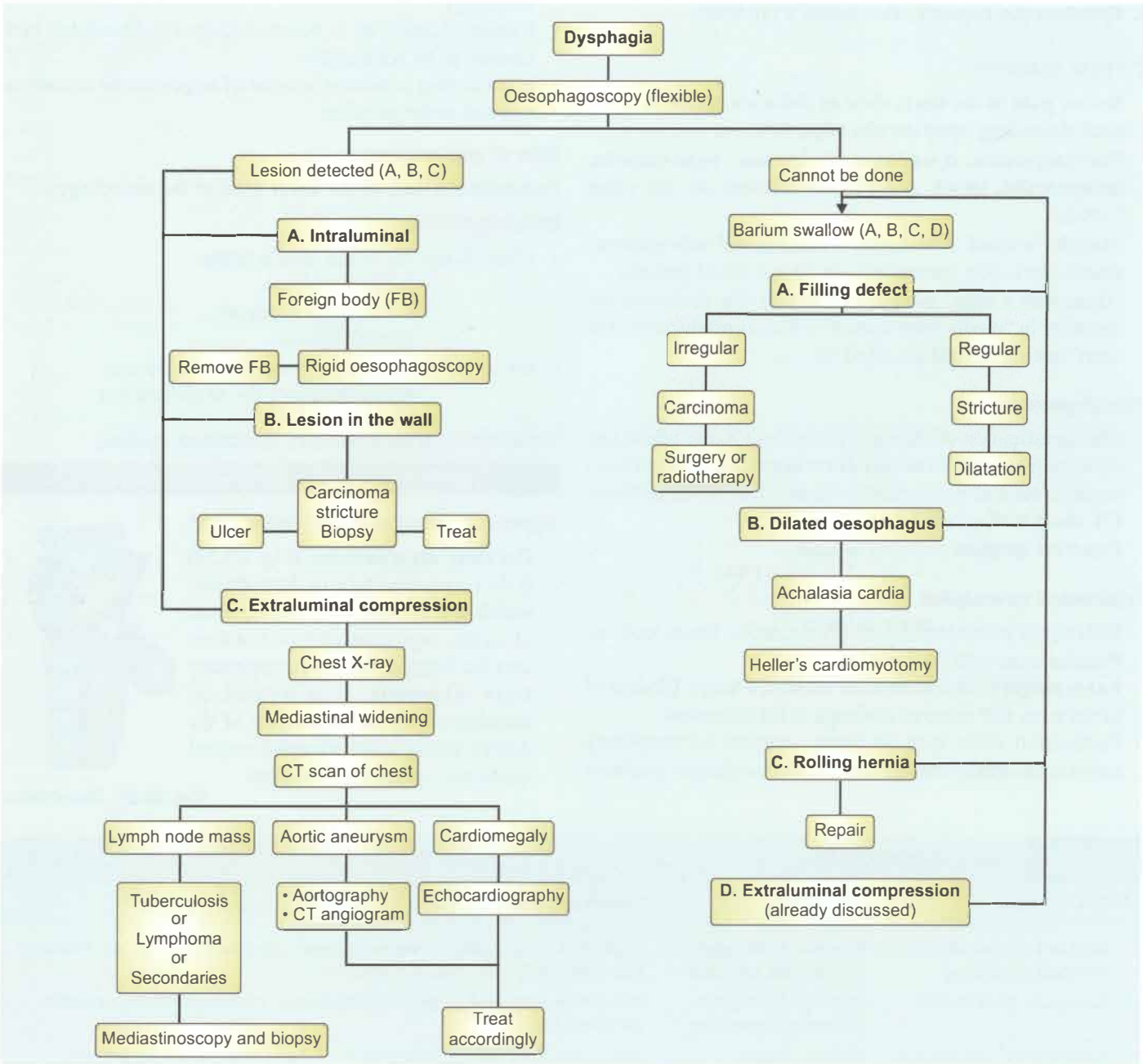


Fig. 22.56: Investigations of a case of dysphagia

PEARLS OF WISDOM

In the absence of usual causes mentioned above, acute dysphagia in an old man may be a manifestation of vertebrobasilar insufficiency.

2. **Chronic dysphagia:** Stricture, achalasia, Plummer-Vinson syndrome and carcinomas produce chronic dysphagia. The increasing difficulty to swallow, first to solids and later to liquids is typical of carcinoma oesophagus (in achalasia cardia, it is the reverse).
3. **Age and sex:** Achalasia and Plummer-Vinson's syndrome is common in females, carcinoma in men and foreign body in children.
4. **Change in voice** or even hoarseness with dysphagia suggests advanced laryngeal carcinoma.

PEARLS OF WISDOM

Dysphagia means difficulty in swallowing. Odynophagia means painful swallowing as in quinsy, retropharyngeal abscess, tonsillitis, etc.

RARE CAUSES OF DYSPHAGIA (RINGS, SLINGS AND WEBS)

1. **Vascular slings and pulmonary artery slings:** Occur due to developmental anomalies of great vessels causing dysphagia. More common is aortic arch anomaly wherein the right subclavian artery arising from the descending aorta travels behind oesophagus creating incomplete vascular ring to complete the course. Pulmonary artery sling is due to left pulmonary artery arising from right pulmonary artery causing anterior compression of oesophagus.
2. **Oesophageal ring: Schatzki's ring:** It is located at the squamo-columnar junction with oesophageal squamous epithelium above and columnar epithelium below. There is a concentric symmetrical narrowing with restricted distensibility of lower oesophagus. It presents as dysphagia and episodic aphagia. Diagnosis is by barium oesophagography. It is treated by repeated bougie dilatation. The cause is obscure but there is strong association with reflux disease. Many rings are incidental findings in radiology. They do not have any symptoms.
3. **Webs:** Oesophageal webs are uncommon causes of dysphagia. They are treated by oesophageal dilatation. *See* clinical notes.

CLINICAL NOTES

A 60-year-old agriculturist was referred to the department of ENT for dysphagia of 3–4 days duration. A Registrar who saw the case did rigid oesophagoscopy under GA. The findings were normal. Later in the evening he was called to see this patient who had rigid abdomen. A general surgeon was consulted. He suspected a perforation. An X-ray abdomen (erect) however, was normal. A senior faculty surgeon was consulted who examined the case properly and gave a correct diagnosis. It was a case of tetanus with mild trismus. The patient had injured his left thumb a few days back. The anaesthesiologist acknowledged later that there was some difficulty in opening the patients mouth during endotracheal intuabtion.

Occasionally, patients undergo gastroscopy for dysphagia which will be normal only to realise later that what he is having is a stroke!!!.

SURGICAL ANATOMY OF THE DIAPHRAGM

- The diaphragm is a dome-shaped thin sheet of muscle which separates the thoracic cavity from the abdominal cavity.
- It is derived from the innermost layer of the muscles of the body. Hence, it arises in continuity with transversus abdominis fibres from within the costal margins.
- It has a right and left dome with a central tendon (Table 22.9).

Attachments

- Anteriorly: Lower sternum
- Laterally: Costal margin
- Posteriorly (crura): First three lumbar vertebrae

Table 22.9 Diaphragmatic hiatus (Fig. 22.57)

Hiatus	Location	Structures passing through
1. Aortic hiatus (median arcuate ligament)	Posteriorly opposite 12th thoracic vertebra	Aorta, thoracic duct, azygos vein
2. Oesophageal hiatus (formed by right crus)	Anterior at the level of 10th thoracic vertebra 1 inch to the left side	Oesophagus, vagus nerves, oesophageal branches of left gastric artery, veins and lymphatics
3. Caval opening	Just to the right of the midline, opposite 8th thoracic vertebra	Inferior vena cava, right phrenic nerve

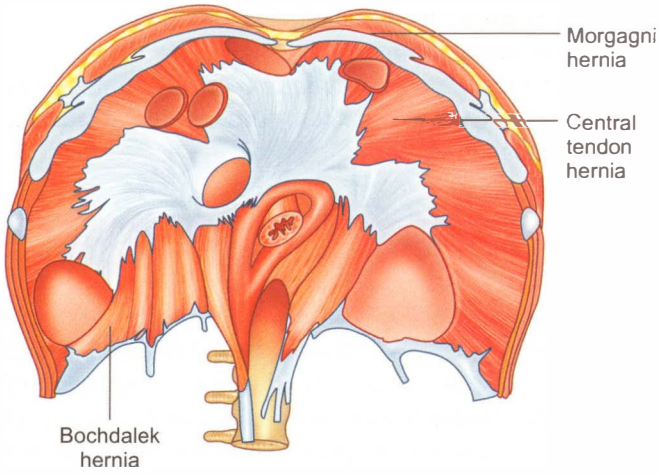


Fig. 22.57: Diaphragmatic hiatus

Fig. 22.58: Bochdalek hernia showing intestine above the liver as soon as laparotomy is done. The defect was two fingers (5 cm) in diameter. Nonabsorbable sutures were used to close the defect (*Courtesy:* Dr Balakrishna Shetty, Surgeon, Chinmaya Hospital, Kundapur)

Nerve supply

The muscles of the dome on its abdominal surface is supplied by phrenic nerve (C4). The crura are supplied by the lower intercostal nerves.

Blood supply

Lower five intercostal arteries, subcostal arteries, and right and left phrenic arteries supply diaphragm.

DIAPHRAGMATIC HERNIA

- Maldevelopment of the septum transversum results in diaphragmatic hernia. The causes of diaphragmatic hernia are of two types: Congenital and acquired. The acquired diaphragmatic hernias are very often (almost always) due to road traffic accidents or due to stab injuries of the abdomen. They are discussed in Chapter 36.
- Majority of congenital hernias present shortly after birth. However, a few cases present late to the hospital, and most of such children are treated for recurrent respiratory tract infection elsewhere, without a proper diagnosis.
- 80% of congenital diaphragmatic hernias occur on the **left side.**

Sites of congenital diaphragmatic hernia

1. Hernia through **the foramen of Bochdalek:** This is the most common diaphragmatic hernia in children. These hernias occur through the persistence of the pleuroperitoneal canals. Respiratory distress, shift of mediastinum and scaphoid abdomen clinch the diagnosis (Fig. 22.58).
2. Hernia through the **foramen of Morgagni (parasternal hernia):** These hernias present in late childhood or in adult life with features of partial (subacute) intestinal obstruction, pain in the right hypochondrium or tightness in the chest (Fig. 22.59). This is because majority of them contain transverse colon.

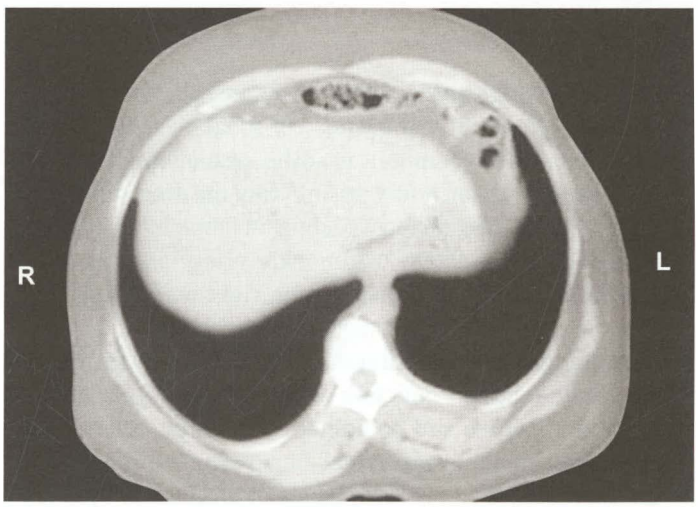


Fig. 22.59: Morgagni hernia—CT scan showing intestine (colon within the thorax) (*Courtesy:* Dr Srinivas Pai, Dr Nitin Nagpal, Dr Rohith Jain, Dept of Surgery, KMC, Manipal)

3. Herniation through the **central tendon:**
   - This can affect the right or the left apex of the cupola or the central portion. The contents may be fundus of the stomach on the left side and a portion of the liver on the right side.
   - These cases are very often diagnosed accidentally by routine chest radiograph.
4. **Eventration:** Due to paralysis or atrophy of the muscle fibres, one or both hemidiaphragms are weak and they are elevated in position. Truly speaking, this is not a hernia, but signs and symptoms are almost like other types of hernia.

Diagnosis

- Chest radiograph: Gas and fluid level is within the chest.
- The thin rim of the diaphragm, as seen in a plain radiograph, is broken or shows a defect.
- CT chest and abdomen is the gold standard investigation.

Treatment

- The repair of the defect is done by using nonabsorbable sutures.
- Eventration of the diaphragm is repaired by plicating the redundant diaphragm.

TRACHEO-OESOPHAGEAL FISTULA

Types

In majority of the cases (85%), lower end of the oesophagus communicates with the trachea and upper end is blind (Fig. 22.60).

Associated anomalies (Key Box 22.19)

KEY BOX 22.19

ASSOCIATED ANOMALIES

V : Vertebral defects
A : Anorectal malformation
C : Cardiac defect (PDA / VSD)
TE : Tracheo-oesophageal fistula
R : Radial hypoplasia and renal agenesis

Clinical features

- Continuous pouring of saliva from the mouth in the newborn baby is a diagnostic feature.
- Newborn baby regurgitates all feeds.
- Aspiration, cough and cyanosis are other features.
- It is commonly associated with maternal hydramnios (50%).
- TOF should be recognised and diagnosed within 24 hours of birth—by introducing a red rubber catheter.

Management

- Right thoracotomy, ligation of TOF and primary anastomosis of oesophageal ends.
- Feeding gastrostomy and oesophagostomy in type III fistula.
- In difficult cases, where there is a long atretic segment, colonic or gastric transposition is required.

Complications

Pneumonia, reflux, dysphagia, anastomotic leak.

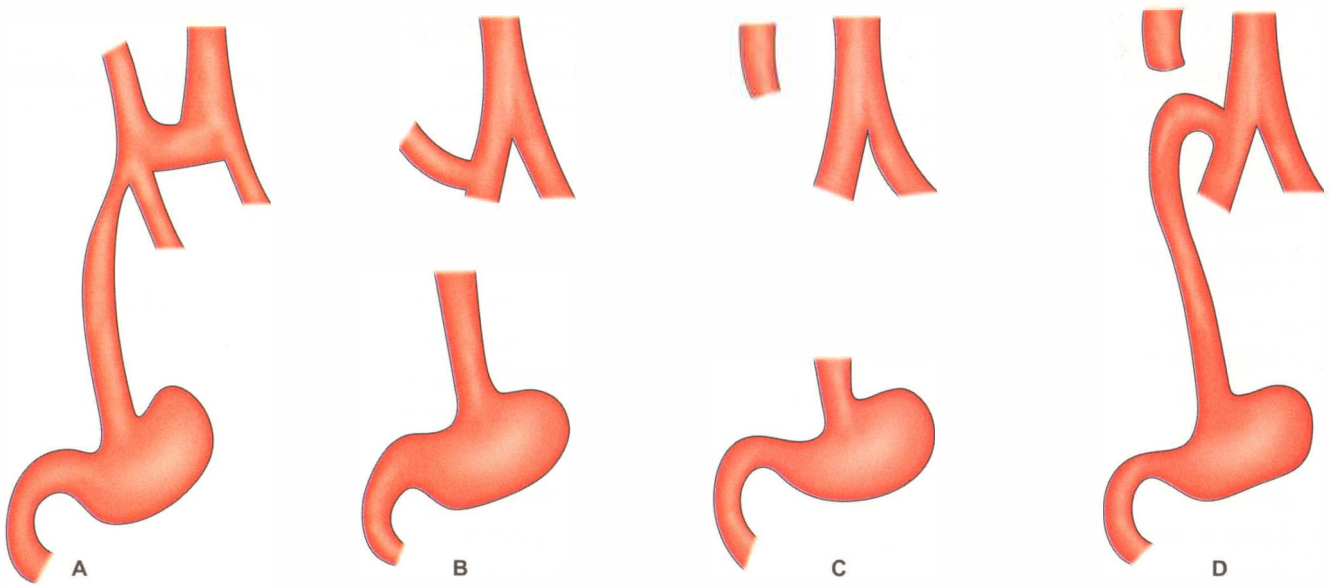
MISCELLANEOUS

See the following two Key Boxes 22.20 and 22.21.

KEY BOX 22.20

INVESTIGATION OF CHOICE

- Investigation of choice for diagnosis with histological proof is **endoscopic biopsy**.
- Investigation of choice to identify haematogenous metastasis is **CT scan**.
- Investigation of choice to detect tracheal/bronchial involvement is **bronchoscopy**.
- Investigation of choice to know the depth of invasion is **endosonography**.
- Investigation of choice to detect enlarged nodes which are malignant or nonmalignant is **PET scan**.



Figs 22.60A to D: Types of tracheo-oesophageal fistula: (A) H-type, (B) Lower end is blind and upper end is connected to trachea, (C) Both ends are blind and (D) Upper blind end and lower end is connected to trachea—the most common variety (85%)

KEY BOX 22.21

INTERESTING 'MOST COMMON' FOR OESOPHAGUS

- Most common among the oesophageal diverticula is Zenker's diverticulum.
- Most common benign oesophageal tumour is leiomyoma or stromal tumour (GIST).
- Most common upper GI disorder of the Western world is gastro-oesophageal reflux disease.
- Most common histological type of carcinoma oesophagus in the world is squamous cell carcinoma.
- Most common histological type of carcinoma oesophagus among whites is adenocarcinoma.
- Most commonly adenocarcinoma affects lower oesophagus.
- Most common presenting feature of Barrett's oesophagus is dysphagia.
- Most common site of gastrointestinal involvement in scleroderma is smooth muscle of oesophagus.
- Most common cause of oesophageal perforation is instrumentation.
- Most serious but rare type of perforation is Boerhaave syndrome—barotrauma, seen in gluttons.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been updated.
- Mechanism of swallowing and surgical anatomy correlated to surgery has been discussed.
- All topics have been edited with colour pictures, new key boxes and tables.
- **Endoscopic resection:** It is attempted as a cure for carcinoma confined to mucosa. It is guided by endosonography. Re-epithelialisation is complete by 3 weeks.
- Achalasia and carcinoma oesophagus have been updated.
- POEM—per oral endoscopic myotomy has been added.
- 'GORD' has been refined.

MULTIPLE CHOICE QUESTIONS

1. Following are disorders of the pharyngo-oesophageal junction *except:*

A. Stroke
B. Myasthenia
C. Cricopharyngeal achalasia
D. Nutcracker oesophagus

2. Following are true for oesophageal lymphatics *except*:

A. Extensive lymphatic plexus in the submucosa
B. They are classified as perioesophageal, paraoesophageal and lateral oesophageal lymph nodes
C. Presence of coeliac nodes indicate inoperability
D. Longitudinal lymphatics are 6 times less than transverse

3. Which one of the following is highest percentage of premalignant condition for carcinoma oesophagus?

A. Tylosis
B. Lye strictures
C. Achalasia
D. Oesophagus

4. Factors for adenocarcinoma oesophagus are following *except:*

A. Barrett's oesophagus
B. Obesity
C. Reflux oesophagitis
D. Oesophageal web

5. Factors for development of reflux disease of oesophagus include following *except*:

A. Obesity
B. Absence of intra-abdominal length of oesophagus
C. *Helicobacter pylori* infection
D. Defective angle of His

6. Clinical features of reflux disease of oesophagus include following *except:*

A. Heartburn
B. Vomiting
C. Epigastric pain
D. Regurgitation

7. Gold standard for the treatment of reflux disease is:

A. Barium meal
B. Oesophagoscopy
C. 24-hour pH monitoring
D. Endosonogram

8. The most effective drugs for reflux disease are:

A. Alginates
B. Antacids
C. H_2 receptor antagonists
D. Proton pump inhibitors

9. The most effective curative treatment for carcinoma oesophagus is:
A. Radiotherapy
B. Oesophagectomy
C. Photodynamic therapy
D. Chemotherapy

10. Following are features of Barrett's oesophagus *except:*
A. 3 cm or more of columnar epithelium
B. Cardiac metaplasia
C. Development of squamous cell carcinoma
D. Presence of mucus secreting goblet cells (intestinal metaplasia)

11. Which of the following conditions predisposes to volvulus of the stomach?
A. Sliding hernia
B. Para-oesophageal hernia
C. Following gastrojejunostomy
D. Wide hiatus

12. Following are true for PET scan *except:*
A. Drug used is oral flurodeoxycolic acid
B. Combining PET with CT is better for diagnosis
C. Can be used after chemotherapy to see the response of the tumour
D. High metabolic activity—glycolytic pathway

13. Following are true for transhiatal oesophagectomy *except:*
A. Thoracotomy is a must for completion of the procedure
B. Anastomosis is in the neck
C. Ideal for lower oesophageal cancers
D. Upper oesophagus mobilisation is blind

14. About achalasia cardia following are true *except:*
A. It affects lower oesophageal end
B. It is due to loss of inhibitory neurons
C. Dilated oesophagus above contain normal ganglion cells
D. In vigorous achalasia normal ganglion cells are present

15. About treatment of achalasia cardia following are true *except:*
A. Balloon dilatation is ideal for patients above the age of 45 years
B. Botulinum toxin injection gives the permanent relief from dysphagia
C. Sublingual nifedipine can relieve symptoms
D. Myotomy with anterior fundoplication is a good surgical procedure

16. Schatzki's ring is associated with:
A. Proximal oesophagus
B. Carcinoma oesophagus
C. Reflux oesophagitis
D. Congenital ring

17. For normal swallowing stricture should be dilated to at least.....
A. 12 mm diameter B. 14 mm diameter
C. 16 mm diameter D. 18 mm diameter

18. Following are true for Mallory-Weiss syndrome *except:*
A. The vertical split occurs in the lower end of oesophagus in majority of cases
B. Surgery is rarely required
C. Endoscopic sclerotherapy is very useful treatment
D. More often it is a mucosal tear than rupture

19. For the diagnosis of intraperitoneal metastasis, which is the ideal investigation?
A. Ultrasound B. CT scan
C. MRI D. Diagnostic laparoscopy

20. Following are true about Heller's cardiomyotomy *except:*
A. It is best done by laparoscopic method
B. Many surgeons add fundoplication
C. Myotomy should be more in length in the stomach side than oesophagus
D. Reflux is a major complication

21. Most accurate method for the diagnosis of T stage for oesophageal cancer is by:
A. Upper gastroduodenal scopy
B. Barium swallow
C. CT scan
D. Endosonogram

ANSWERS

1 D	2 D	3 A	4 D	5 C	6 B	7 B	8 D	9 B	10 C
11 B	12 A	13 A	14 C	15 B	16 C	17 C	18 A	19 D	20 C
21 D									

23

Stomach and Duodenum

- Surgical anatomy
- Gastric physiology
- *H. pylori* infection
- Gastritis
- Peptic ulcer disease
- Acute complications of peptic ulcer
- Chronic complications of peptic ulcer
- Carcinoma stomach
- Gastrointestinal stromal tumours (GISTs)
- Gastric lymphoma
- Complications of gastrectomy
- Acid function tests
- Acute dilatation of the stomach
- Volvulus of the stomach
- Bezoars
- Idiopathic hypertrophic pyloric stenosis
- Chronic duodenal ileus
- Duodenal anatomy and obstruction
- Gastric surgery for morbid obesity
- What is new?/Recent advances

Introduction

The oesophagus continues from the oesophagogastric (OG) junction as a muscular tube—the stomach.

SURGICAL ANATOMY

Fundus

Part of the stomach which projects upwards and lies in contact with the left dome of diaphragm. It is usually full of gas.

Significance

- To identify the side (right or left) of the body in a plain X-ray abdomen.
- In achalasia cardia, fundic air bubble is absent.
- Fundic **'Wrap'** is used in hiatus hernia.
- During mobilisation of the fundus as in splenectomy or other upper gastric surgery, short gastric arteries need to be divided. If ligatures are too close to the stomach near the fundus, gastric fistula may occur due to **necrosis of the stomach**.
- GISTs (gastroinstestinal stromal tumours) are common in fundus.

Body

Extends from fundus to *incisura angularis*. It has a lesser curvature and a greater curvature.

Significance

- Ability to have a large meal is due to receptive relaxation of the body of the stomach.
- Greater curvature is located at the level of umbilicus.
- Classical gastrojejunostomy (GJ), anterior or posterior, involves using body of the stomach.
- Posteriorly, it is related to the lesser sac and pancreas. Carcinoma of the body may infiltrate pancreas—necessitates careful dissection to separate from pancreas (sometimes not resectable).

Pyloric antrum

It extends from incisura till pylorus. Pylorus is thicker than the rest of the stomach. It is a sphincter of circular muscle fibres. Its canal is usually closed.

Significance (Table 23.1)

- Pyloric antrum is a common site for gastritis, ulcer and carcinoma.
- Incompetence of pyloric sphincter results in severe duodenogastric reflux.
- It is in close contact with the head of pancreas. During gastrectomy, extreme care has to be taken to mobilise the antrum to avoid bleeding in the pancreatic head region.

Greater curvature

It lies in contact with transverse colon and gastrocolic omentum (Fig. 23.1 and Key Box 23.1). This has to be divided from transverse colon during gastrectomy which is done for carcinoma or ulcer.

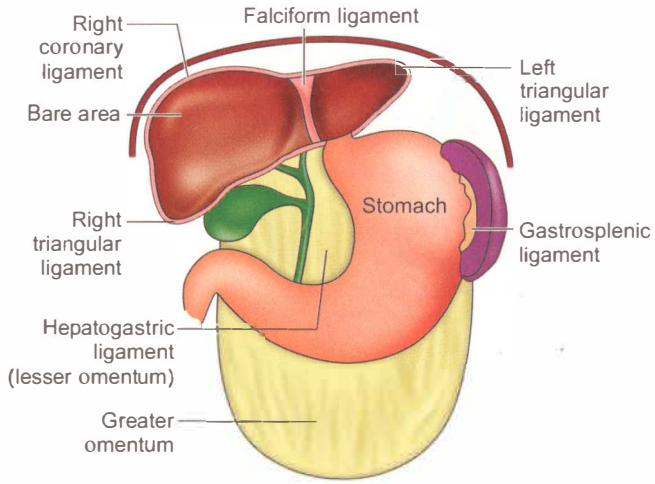


Fig. 23.1: Stomach, omentum and ligaments

Lesser omentum

- The lesser omentum is a double-layered structure.
- It is suspended between the lesser curvature of the stomach and the proximal 0.5 inch (2 cm) of the first part of the duodenum inferiorly and the porta hepatis and the fissure of the ligamentum venosum superiorly.

The lesser omentum is divided into two ligaments:

1. **Hepatogastric**
2. **Hepatoduodenal:** Located within the lesser omentum are the **hepatic triad**, branches of the anterior vagus nerve, some lymph nodes, and the right and left gastric arteries.

Vagus nerves

- The left and right vagus nerves descend parallel to the oesophagus and form oesophageal vagal plexus between the level of the tracheal bifurcation and level of the diaphragm.
- From this plexus, two vagal trunks, anterior and posterior form and pass through the oesophageal hiatus of the diaphragm **(mnemonic LARP: Left trunk—anterior gastric wall; right trunk—posterior gastric wall).**

KEY BOX 23.1

GREATER OMENTUM IS A FRIEND AND SOMETIMES ENEMY OF THE PATIENT AND THE SURGEON

- The greater omentum is the **policeman** of the peritoneal cavity. It exhibits **protective action** in inflammatory processes and perforations.
- **Omentoplasty** (omental wrapping of anastomosis) may decrease the occurrence of anastomotic leakage when oesophagogastric anastomosis is performed for cancer of oesophagus or as in closure of duodenal perforation.
- It has many **important applications** and can be used in colorectal surgery, reconstruction of the irradiated pharyngeal wound and packing of bleeding liver wounds.
- It was used to **treat peripheral vascular disease** such as TAO wherein direct arterial surgery was not possible.
- It becomes an enemy when involved with metastasis or directly by malignant neoplastic process from ovary or colorectal cancer. Omentum is one of the common sites of involvement in tuberculosis also. Omentum itself can act like a **band** and can cause intestinal obstruction.

Blood supply of the stomach (Fig. 23.2)

It is mainly supplied by coeliac trunk and its branches:

1. **Left gastric artery** is a direct branch of coeliac trunk. It ascends up to oesophageal hiatus and turns to the right along the lesser curvature of stomach. It branches and anastomoses with branches of right gastric artery and supplies anterior and posterior wall of the stomach. There is true anastomosis between branches of left gastric artery and branches from other arteries.

Table 23.1 Anatomy and surgical significance

2 Orifices	Recognition	Pathology	Result
• Pyloric orifice	Recognised by prepyloric vein of Mayo	Incompetence causes duodeno-gastric reflux	Biliary gastritis
• Cardiac orifice	Lower end of oesophagus	Incompetence causes acid reflux into lower end of oesophagus	Barrett's oesophagus
2 Curvatures			
• Lesser curvature	Concave lesser omentum is attached to it	Site of benign gastric ulcer	Hour-glass contracture
• Greater curvature	Convex greater omentum is attached to it	Site of carcinomatous ulcer	Carcinoma stomach
Part	**Common pathology**	**Commonly used for**	**Nature of the condition**
• Fundus	GIST	Fundic wrap	Hiatus hernia
• Body	Carcinoma	Gastrojejunostomy	Pyloric stenosis
• Pylorus	Pyloric stenosis, carcinoma	Pyloric myotomy in idiopathic pyloric stenosis	

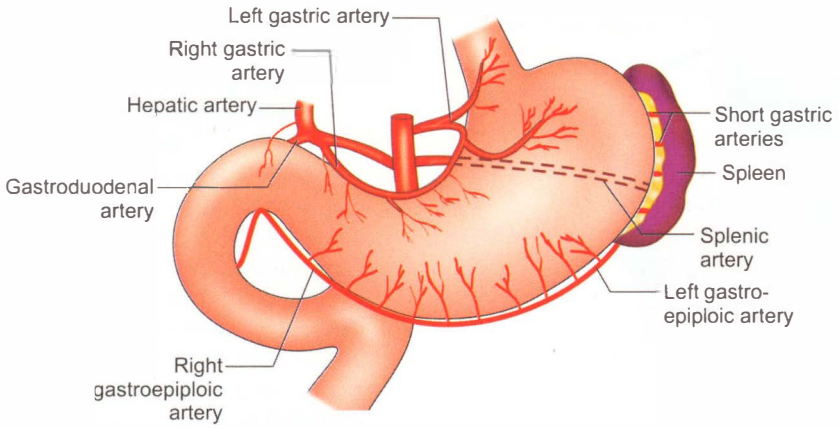


Fig. 23.2: Blood supply of the stomach

2. **Right gastric artery** is a branch of hepatic artery which comes from coeliac trunk. It also supplies lesser curvature and body of stomach, along with left gastric artery.
3. **Left gastroepiploic artery** arises from splenic artery and supplies greater curvature of stomach and anastomoses with right gastroepiploic artery.
4. **Right gastroepiploic artery** is a branch of gastroduodenal artery, which is a branch of hepatic artery.
5. **Short gastric arteries** are the branches of splenic artery. They supply the fundus of the stomach. They are also called *vasa braevia*.

Venous drainage

- Veins run with the corresponding arteries.
- Right and left gastric veins drain into the portal vein directly.
- Right gastroepiploic vein joins superior mesenteric vein.
- Left gastroepiploic vein and *vasa braevia* join splenic vein
- Prepyloric vein of Mayo is a useful guide to the junction between stomach and duodenum.

Surgical importance

- Because of extensive anastomoses of blood vessels, (extramural and intramural collateral vessels) stomach can survive with right gastric and right gastroepiploic arteries only. Thus stomach can be used to replace the entire oesophagus after oesophagectomy—gastric pull up.
- The order of ligation of blood vessels in gastrectomy is as follows: Left gastroepiploic, right gastroepiploic, right gastric (then stomach is divided) and lastly, left gastric artery.
- Gastroduodenal artery, a branch of hepatic artery runs behind first part of duodenum and divides into right gastroepiploic artery and superior pancreaticoduodenal artery.
- It is this artery which bleeds when a posterior duodenal ulcer erodes into it.

Lymphatic drainage (Fig. 23.3)

It is an important pathway for spread of carcinoma of the stomach. The spread occurs both by emboli and permeation.

1. Right gastric nodes/suprapyloric nodes, mainly drain the pyloric antrum.

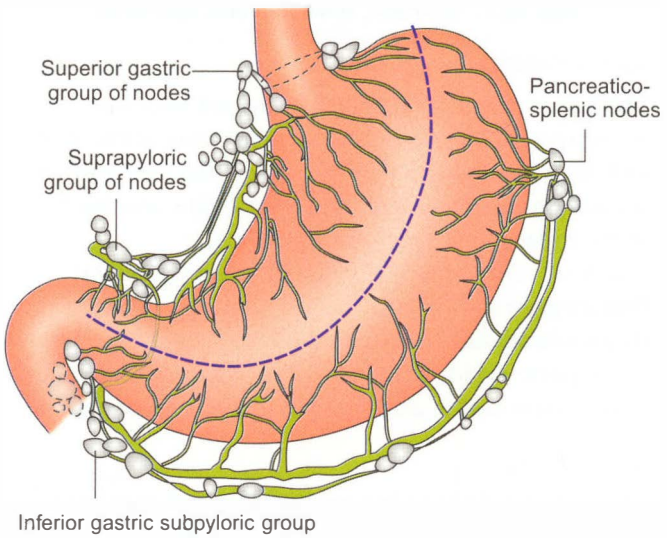


Fig. 23.3: Lymphatic drainage of the stomach

2. Subpyloric nodes/gastroepiploic nodes (right) drain the greater curvature of stomach and pyloric antrum.
3. Left gastroepiploic nodes (splenic nodes) drain the upper portion of stomach, mainly the fundus (carcinoma of fundus).
4. Left gastric (superior gastric) nodes drain the lesser curvature and body of the stomach (anterior and posterior wall).
5. Coeliac nodes receive lymph from the entire foregut (including stomach) and drain directly into the *cisterna chyli* and the thoracic duct. Later, mediastinal nodes and left supraclavicular nodes (**Virchow nodes**) are involved.

Lymphatic zones are discussed under carcinoma stomach.

Lymphatics of the stomach

The lymphatics of the stomach are formed from two networks: Intrinsic and extrinsic.

Intrinsic network: The submucosal lymphatic plexus is perhaps the richest in anastomoses.

- Upward invasive spread of gastric cancer to the oesophagus is common but downward mucosal or submucosal invasion of the duodenum is not common.

- Hence, gastrectomy with distal oesophagectomy is done for carcinoma of the proximal stomach.

Extrinsic network: The extrinsic lymphatic vessels of the stomach follow the gastric veins.

- They drain to perigastric lymph nodes, which in turn drain to the lymph nodes located at the coeliac axis.

1. **The superior gastric group** drains lymph from the upper lesser curvature into the left gastric and paracardiac nodes.
2. **The suprapyloric group** of nodes drains the antral segment on the lesser curvature of the stomach into the right suprapancreatic nodes.
3. **The pancreaticolienal group** of nodes drains lymph high on the greater curvature into the left gastroepiploic and splenic nodes.
4. **The inferior gastric and subpyloric group** of nodes drains lymph along the right gastroepiploic vascular pedicle.

- All four zones of lymph nodes drain into the coeliac group and then into the thoracic duct.

GASTRIC PHYSIOLOGY

Hypergastrinaemia

Various causes of surgical interest are given in Key Box 23.2.

KEY BOX 23.2

CAUSES OF HYPERGASTRINAEMIA

Ulcerogenic causes	Nonulcerogenic causes
• Antral G cell hyperplasia or hyperfunction	• Antisecretory agents (PPIs)
• Retained excluded antrum	• Atrophic gastritis
• Zollinger-Ellison syndrome	• Pernicious anaemia
• Gastric outlet obstruction	• Acid-reducing procedure (vagotomy)
• Short-gut syndrome	• *Helicobacter pylori* infection
	• Chronic renal failure

Gastric acid secretion (Table 23.2)

- Gastric acid secretion by the parietal cells is regulated by three local stimuli:
  1. Acetylcholine, 2. Gastrin, 3. Histamine.
- These three stimuli account for basal and stimulated gastric acid secretion.
- **Acetylcholine is the principal neurotransmitter** modulating acid secretion and is released from the vagus and parasympathetic ganglion cells.
- Vagal fibres innervate not only parietal cells but also G cells and enterochromaffin-like (ECL) cells to modulate release of their peptides.
- **Gastrin has hormonal effects on the parietal cell** and stimulates histamine release (Fig. 23.4).
- **Histamine has paracrine-like effects** on the **parietal cell**. Its release from ECL cells plays a central role in the regulation of acid secretion by the parietal cell. Ingestion of a meal stimulates vagal fibres to release acetylcholine ***(cephalic phase)***. Binding of acetylcholine to parietal cell and G cell results in the release of histamine, hydrochloric acid and gastrin, respectively. Acetylcholine also inhibits somatostatin release by inhibiting ECL.
- Following a meal, G cells are also stimulated to release gastrin, which interacts with receptors located on ECL cells and parietal cells to cause the release of histamine and hydrochloric acid ***(gastric phase)***. Release of somatostatin from D cells decreases histamine release and gastrin release from ECL cells and G cells, respectively. In addition, somatostatin inhibits parietal cell acid secretion (not shown). The principal stimulus for activation of D cells is **antral luminal acidification** (not shown).
- As depicted, somatostatin exerts inhibitory actions on gastric acid secretion.
- Release of **somatostatin from antral D cells** is stimulated in the presence of intraluminal acid to a pH of 3 or less.
- After its release, somatostatin inhibits gastrin release through paracrine effects and also modifies histamine release from ECL cells.

Table 23.2 Gastric secretion

	Cells	Location	Function/mediators
1. Acid secretion 150–160 mEq/L	• Oxyntic glands, principal cell type is parietal	• Fundus and body	• Cephalic—vagus • Gastric—gastrin
2. Mucus	• Columnar epithelium	• Entire stomach	• Intestinal—not clear • Luminal cytoprotection from acid, pepsin and ingested substances
3. Pepsinogen I and II	• Chief cells—located in deeper areas of oxyntic gland	• Body and fundus	• Vagal stimulation
4. Mucus, gastrin	• Antrum	• Antrum	• Cytoprotection
5. Bicarbonate	• Mucus cells	• Stomach	• Vagal stimulation increases HCO_3^- secretion • Prostaglandin E_2
6. Intrinsic factor	• Parietal cells	• Fundus	• Necessary for absorption of B_{12}
7. Peptides, histamine	• Enterochromaffin like (ECL) cells • D cells (delta)	• Fundus • Antrum	• Release of peptides • Somatostatin inhibits acid secretion

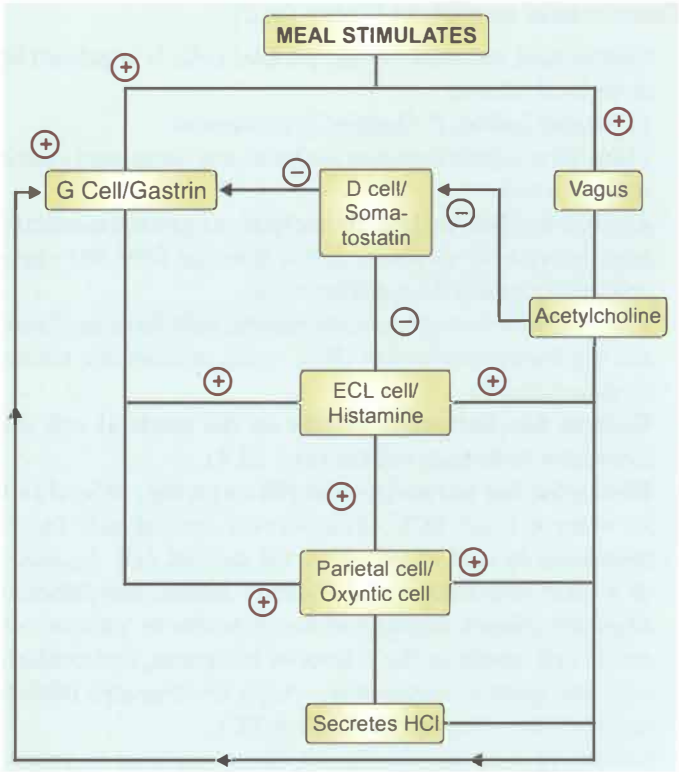


Fig. 23.4: Secretion of acid

- In some patients with peptic ulcer disease (PUD), this negative feedback response is defective.
- Consequently, the precise state of acid secretion by the parietal cell is dependent on the overall influence of the positive and negative stimuli.

Functions of gastric acid (Key Box 23.3)

KEY BOX 23.3

FUNCTIONS OF GASTRIC ACID

- Gastric acid plays a critical role in the digestion of a meal.
- It is required to convert pepsinogen into pepsin, which is necessary for hydrolysis of proteins into polypeptides.
- Gastric acid also elicits the release of secretin from the duodenum, which results in pancreatic bicarbonate secretion.
- In addition, gastric acid functions to limit colonisation of the upper GI tract with bacteria.

Gastric analysis

- There are numerous ways to assess acid secretion in the stomach.
- **Aspiration** of gastric contents through a nasogastric tube is probably the most accurate.
- The study requires complete emptying of gastric contents followed by instillation and recovery of 50 ml of saline.

KEY BOX 23.4

PROMOTILITY AGENTS THAT ACCELERATE GASTRIC EMPTYING

Drug	Mechanism
Metoclopramide	Dopamine antagonist
Domperidone	Dopamine antagonist
Erythromycin	Motilin agonist
Bethanechol	Cholinergic agonist
Neostigmine	Cholinergic agonist

- The stomach is then aspirated every 5 minutes for 1 hour and the aspirates pooled in 15 minute aliquots.
- At the end of 1 hour, the stomach is stimulated to secrete acid by intravenous (IV) administration of a secretogogue such as **Histamine** (2 µg/kg) or **Pentagastrin** (6 µg/kg).
- Aspiration of the stomach continues with four 15-minute collections obtained over a 1-hour period.
- The volume of collections is measured, and each aliquot is titrated to determine the amount of hydrogen ions present.
- The rate of secretion is expressed as the number of milli-equivalents produced per hour during the basal (unstimulated) state and during maximal and peak acid output (Key Box 23.4).
- **Maximal acid output** (MAO) is obtained by averaging the output of the two final 15 minute periods. MAO is in the range of 10 to 15 mEq/hour.
- **Peak acid output** is the highest rate of secretion obtained during a 15-minute period following secretogogue stimulation.
- **Basal acid output (BAO)** is generally about 2 to 3 mEq/hour.

HELICOBACTER PYLORI INFECTION

- It is now established that 90% of duodenal ulcers and 75% of gastric ulcers are associated with *H. pylori* infection.
- **Warren and Marshall** were the first to identify and isolate the organism, originally referred to as ***Campylobacter pyloridis.***
- The organism is a **spiral or helical gram-negative rod** with **4 to 6 flagella** and resides in gastric-type epithelium within or beneath the mucus layer, which protects it from both acid and antibiotics.
- Its shape and flagella aid its movement through the mucus layer, and it produces a variety of enzymes that help it adapt to a hostile environment.
- Most notably, it is one of **the most potent producers of urease among any bacteria yet described.**
- This enzyme is capable of **splitting urea into ammonia and bicarbonate,** creating an alkaline microenvironment in the setting of an acidic gastric milieu, which facilitates establishing a diagnosis of this organism by various laboratory tests.
- The organism is microaerophilic, and the optimal temperature for isolation is 35 to 37°C, with growth occurring after 2 to 5 days.

- Interestingly, *H. pylori* **can only live in gastric epithelium** because only gastric epithelium expresses specific adherence receptors *in vivo* that can be recognised by the organism. Thus, it can also be found in **heterotopic gastric mucosa in proximal oesophagus** and **Barrett's oesophagus,** in gastric metaplasia in the duodenum and Meckel's diverticulum, and in heterotopic gastric mucosa in the rectum.
- **The mechanisms responsible for *H. pylori*-induced GI injury** is not very clear but three potential mechanisms have been proposed:
  1. Production of toxic products to cause local tissue injury
  2. Induction of a local mucosal immune response
  3. Increased gastrin levels with a resultant increase in acid secretion.

Locally produced toxic mediators

1. Urease activity, i.e. ammonia.
2. Cytotoxins.
3. Mucinase that degrades mucus and glycoproteins.
4. Phospholipases that damage epithelial cells and mucus cells.
5. Platelet-activating factor, which is known to cause mucosal injury.

Mediators of *Helicobacter pylori*-induced injury (Key Box 23.5)

- Most *H. pylori* strains express—**vacuolating cytotoxin**.
- **The VacA exotoxin** causes the release of cytochrome c from mitochondria and induces apoptosis.
- *H. pylori* infection is accompanied by an **abnormal T-cell response** which may be partially responsible for the long-term persistence of *H. pylori* infection.

KEY BOX 23.5

MECHANISM OF *HELICOBACTER PYLORI*-INDUCED INJURY—SUMMARY

Helical gram-negative rod

Enzyme produced is urease, which splits urea. Resulting bicarbonate creates alkaline environment. Another enzyme mucinase degrades mucus.

Lives only in gastric epithelium

Injury is due to:

- Toxins such as **vacuolating cytotoxin**
- Increased gastrin levels
- Phospholipases
- Platelet-activating factor
- Increased immunoglobulin response (host response)

Cytotoxins

Other factors

- Local tissue injury
- Local ischaemia

Remember as **HELICO**

Diagnosis (Table 23.3)

H. pylori infection can be diagnosed by both noninvasive and invasive means.

I. Noninvasive methods

- They are **urea breath test, serology, and detection of antigen in stool samples**.
- The urea breath test is based on production of urease by *H. pylori* in the gastric mucosa.
- ^{14}C-labelled urea is ingested and ^{14}C-labelled CO_2 is produced and excreted in the breath.
- This test has a sensitivity and specificity of greater than 90% and indicates ongoing infection.
- The urea breath test is useful for initial diagnosis of infection and for follow-up after eradication therapy.

II. Invasive methods

- The patient should undergo endoscopic diagnosis and it is indicated in following situations.
  - Individuals more than 50 years of age
  - Those with significant symptoms including gastrointestinal bleeding
  - Anaemia
  - Weight loss.
- During endoscopy, antral biopsies can be obtained and the organism cultured in agar containing both urea and a pH-sensitive colorimetric agent.
- *H. pylori* hydrolysis of urea causes a diagnostic change in colour.
- The sensitivity of this test varies from 80 to 100% and specificity exceeds 90%.
- Biopsy also permits histologic examination with visualisation of the organism.

Culture of *H. pylori* is not routine and is usually reserved for recurrent infection and for antibiotic sensitivity testing when second-line therapy has failed.

Treatment

- To be effective, antimicrobial drugs must be combined with gastric acid secretion inhibitors or bismuth salts.
- Cure rates of 80–85% are achieved using combination therapy, usually proton pump inhibitors, ranitidine, or bismuth citrate with two antibiotics.
- The most common antibiotics used are **clarithromycin, amoxicillin, and metronidazole or tinidazole.**
- Metronidazole has been a mainstay of *H. pylori* treatment.
- Eradication therapy with a proton pump inhibitor, metronidazole, and amoxicillin decreases the prevalence of metronidazole-resistant *H. pylori* strains.

Different regimens

Bismuth triple therapy

- Bismuth, 2 tablets four times daily + Metronidazole, 250 mg three times daily + Tetracycline, 500 mg four times daily.

Table 23.3 Test for *Helicobacter pylori*

Test	Indication	Advantages	Disadvantages
1. **Serologic test** (ELISA based)	When the patient has not received treatment for *H. pylori* infection, test of choice is serological test	Noninvasive; sensitivity of > 80%, specificity of about 90%	Does not confirm eradication, because serologic positivity remains for indefinite period after microbiologic cure
2. **Urea breath test**: Based on ability to hydrolyse urea.	This test is to confirm cure of *H. pylori* infection, but no sooner than 4 weeks after completion of therapy	Simple; sensitivity and specificity of 90 to 99%	False-negatives possible if testing is done too soon after treatment for *H. pylori* infection. Small radiation exposure with ^{14}C method, cost and availability are disadvantages
3. **Histologic test** is done by staining with silver, Giemsa, etc.	Presence of *H. pylori* in biopsy material taken during endoscopy establishes the diagnosis	Sensitivity of 80 to 100%, specificity of > 95%; Giemsa stain has sensitivity of > 95% and specificity of 99%	Requires laboratory facilities and experience; when haematoxylin-eosin stain is nondiagnostic, other stains have to be used
4. **Rapid urease test**	Simplest method. However, endoscopy is required.	Simple; rapid (once biopsy specimen has been obtained); sensitivity of 80 to 95%, specificity of 95 to 100%	Invasive; false-negatives possible if testing is done too soon after treatment with proton pump inhibitors, antimicrobials or bismuth compounds
5. **Culture** (not routinely done)	When antimicrobial resistance is suspected after many drug failures	Allows determination of antibiotic susceptibility. Sensitivity is 80% and specificity is 100%	Time-consuming; expensive; usually not necessary unless resistance is suspected

PPI triple therapy

PPI twice daily + Amoxicillin, 1000 mg two times daily + Clarithromycin, 500 mg two times daily or metronidazole, 500 mg two times daily.

Quadruple therapy

PPI twice daily + bismuth, 2 tablets four times daily + metronidazole, 250 mg three times daily + Tetracycline, 500 mg four times daily.

Complications of *Helicobacter pylori* infection

1. Chronic duodenal ulcer
2. Gastric carcinoma
3. Gastro-oesophageal reflux disease
4. Barrett's oesophagus
5. Chronic gastritis
6. Gastric maltoma
7. Oesophageal cancer
8. Idiopathic thrombocytopaenic purpura.

Table 23.4 Gastritis

Type A	Type B	Reflux	Erosive
• Autoimmune—antibodies to parietal cell	• *H. pylori* associated	• After cholecystectomy, gastric surgery	• It occurs due to NSAID
• Fundus and body are affected	• Antrum is affected	• Antrum is affected	• It affects entire stomach
• Atrophy of parietal cell mass. Hence, hypochlorhydria and decreased intrinsic factor	• Predisposes to peptic ulcer disease	• Usually does not give rise to peptic ulcer disease	• It occurs due to defective gastric mucosal barrier
• Hypochlorhydria stimulates antral gastrin. Predisposes to development of gastric cancer	• Can give rise to intestinal metaplasia, dysplasia and predisposes to development of gastric cancer	• Treated by prokinetic agents or bile chelating agents	• Treated by H_2 blockers or proton pump inhibitors

GASTRITIS

- Various forms of gastritis have been depicted in Table 23.4.
- Type B gastritis and erosive gastritis are common.
- Endoscopy and biopsy are the key investigations in all forms of gastritis.
- The treatment depends upon the type of gastritis.
- Rarer forms of gastritis include granulomatous gastritis, lymphocytic gastritis, etc.

PEPTIC ULCER DISEASE

Definition

Acid peptic digestion of the alimentary mucosa resulting in an ulcer is called peptic ulcer disease (PUD). The corrosive effects of acid with proteolytic effect of pepsin are responsible for PUD. Duodenum and stomach are the common sites of peptic ulcer disease. Rarely, they can occur in the jejunum and in Meckel's diverticulum when it contains ectopic gastric mucosa.

Types of peptic ulcer

I. Depending on the site

A. Chronic duodenal ulcer: Typically occurs in the first inch of the first part of the duodenum.

B. Chronic gastric ulcer: Occurs in the lesser curvature adjacent to acid secreting parietal cell mass.

C. Combined: Gastric ulcer type II

- Zollinger-Ellison syndrome

D. Anastomotic ulcer

II. Depending on the duration

A. Chronic peptic ulcer, and B. Acute peptic ulcer

Before we start discussion on peptic ulcer disease, we will discuss nonulcerative dyspepsia (commonly mistaken as peptic ulcer disease) and Zollinger-Ellison's syndrome which is one of the rare causes of peptic ulceration.

NONULCERATIVE DYSPEPSIA

- Dyspepsia is characterised by symptoms related to upper abdominal viscera. Symptoms may include abdominal pain, bloating, indigestion, nausea and heart burn. Heart burn as a single symptom suggests gastro-oesophageal reflux disease and excludes the diagnosis of dyspepsia.
- Nonulcerative dyspepsia is considered when no anatomic or biochemical abnormality is discovered to explain the patient's symptoms.
- After *H. pylori* infection, ingestion of nonsteroidal anti-inflammatory drugs (NSAIDs) is the most common cause of nonulcerative dyspepsia and PUD.
- *H. pylori* infection is always associated with histologic gastritis which is absent in cases of nonulcerative dyspepsia.
- Hence *H. pylori* eradication therapy is not recommended in the treatment of nonulcerative dyspepsia.
- For surgeons, the importance of nonulcerative dyspepsia relates to its place in the differential diagnosis of epigastric pain. There is no role for surgery in the treatment of this disorder.
- Some of these patients who have been diagnosed as nonulcerative dyspepsia may have gall stones or oesophageal reflux disease. Hence abdominal ultrasound and oesophageal manometry is required to rule out these diseases.
- Oesophagogastroduodenoscopy should be done in all the patients with nonulcerative dyspepsia followed by biopsy from stomach to rule out *H. pylori* infection.One important cause of peptic ulcer disease—Zollinger-Ellison syndrome is being discussed first.

ZOLLINGER-ELLISON SYNDROME

Introduction

- Zollinger-Ellison syndrome (ZES) is a **clinical triad consisting of gastric acid hypersecretion, severe PUD, and non-islet cell tumours of the pancreas.**
- The tumours are known to produce gastrin and are referred to as gastrinomas (Fig. 23.5).
- These tumours are usually localised to the head of the pancreas, duodenal wall or regional lymph nodes.
- About one-half of these gastric tumours are multiple and two-thirds are malignant.

Association with MEN

About one-fourth are associated with **multiple endocrine neoplasia syndrome (MEN 1)**. Other tumours in MEN 1 are pituitary adenoma and parathyroid adenoma.

Pathophysiology

Pathophysiologic features of ZES that distinguish it from duodenal ulcer are present and are all explained either by the actions of **gastrin to stimulate gastric acid secretion** and **mucosal growth** or by the action of other associated hormones in the MEN 1 syndrome.

Clinical features

- Abdominal pain and PUD are the hallmarks of the syndrome and typically occur in more than 80% of patients.
- About one-half of patients have diarrhoea secondary to increased gastric acid secretion.
- Weight loss and steatorrhoea also occur secondary to decreased duodenal and jejunal pH and inactivation of lipase.
- Oesophagitis from gastro-oesophageal reflux is also common.

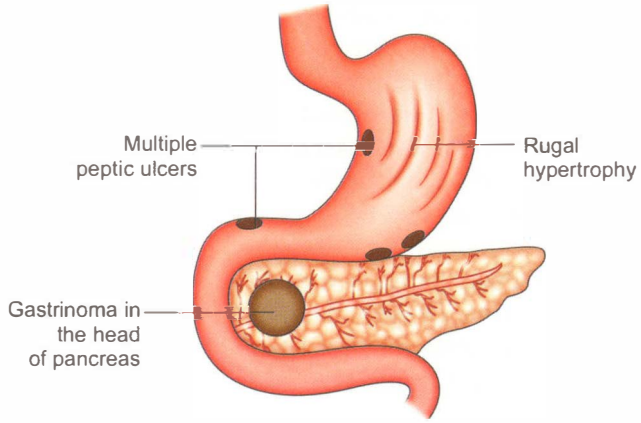


Fig. 23.5: Pictorial representation of gastrinoma (refer page 596 also)

Investigations

I. Endoscopy

- **Demonstrates prominent gastric rugal folds, reflecting the trophic effect of hypergastrinaemia on the gastric fundus in addition to evidence of PUD.**
- Gastrinoma and ZES are always considered and ruled out in patients who have:
  1. Recurrent or intractable PUD despite eradication of *H. pylori* and appropriate antisecretory therapy.
  2. Multiple or atypically located ulcers.
  3. PUD associated with significant diarrhoea.
  4. PUD associated with symptoms of MEN 1 such as hyperparathyroidism or kindred of MEN 1.
  5. Large gastric rugae on endoscopy.
  6. **In those patients with other pancreatic endocrine tumours.**
  7. Patients undergoing elective surgical intervention for PUD have the possibility of gastrinoma included in their preoperative evaluation.

II. Serum gastrin levels

Most patients with gastrinoma have **elevated fasting levels (> 200 pg/ml), and values greater than 1000 pg/ml may be diagnostic.**

III. The secretin test

- It is the most sensitive and specific provocative test for gastrinoma and aids in the differentiation between gastrinomas and other causes of ulcerogenic hypergastrinaemia.
- Secretin (2 μg/kg) is administered intravenously and serum gastrin samples are measured before and after secretin administration at 5-minute interval for 30 minutes.
- An increase in serum gastrin of greater than 200 pg/ml above basal levels is specific for gastrinoma.

Treatment

- Acid suppression therapy with a proton pump inhibitor.
- Localisation of the gastrinoma is performed before operative intervention is made.
- **Noninvasive methods** include computed tomography (CT scanning, MRI, endoscopic ultrasound, and ^{111}In-octreotide scintigraphy (somatostatin receptor imaging).
- Invasive modalities include selective visceral angiography percutaneous transhepatic portal venous sampling for gastrin and selective arterial secretin stimulation test.
- In patients with **resectable gastrinomas**, surgical resection is performed that includes tumour resection from the duodenum, pancreas and regional lymph nodes.
- **Total gastrectomy is rarely indicated** and is reserved for patients who are noncompliant with acid suppression therapy or when the tumour cannot be localised.

CHRONIC PEPTIC ULCER—PEPTIC ULCER DISEASE (PUD)—AETIOLOGY

CHRONIC DUODENAL ULCER (CDU)

Hyperacidity is the chief cause of duodenal ulcer. **No acid, no ulcer** still holds good for CDU.

1. **Neurological causes:** Stimulation of vagus increases secretion of acids. This is brought about by **anxiety, worry, hurry and curry**.
2. **Nonsteroidal anti-inflammatory drugs:** They are responsible for gastric ulcer rather than duodenal ulcer by altering mucosal defence.
3. **Genetic causes:** Family history of duodenal ulcer may be present in a few cases which suggests a genetic cause.
   - Patients with blood group **'O'** are more prone, for the development of CDU because of **increased parietal cell population.**
4. **Food habits:** Spicy food, diet poor in vitamins, smoking and alcohol, alone or in combination precipitate the development of chronic duodenal ulcer.
5. **Bacteriological causes** (Fig. 23.6)
   - ***Helicobacter pylori***, a spirochaetal bacteria has been demonstrated in the submucosa of the antrum and duodenum, from the biopsies of the ulcer. **It increases pH levels by splitting urea and releasing ammonia.** Rise in pH results in proliferation of bacteria.
6. **Endocrinal causes**
   - Zollinger-Ellison syndrome is a non-β cell tumour of the pancreas with hypergastrinaemia.

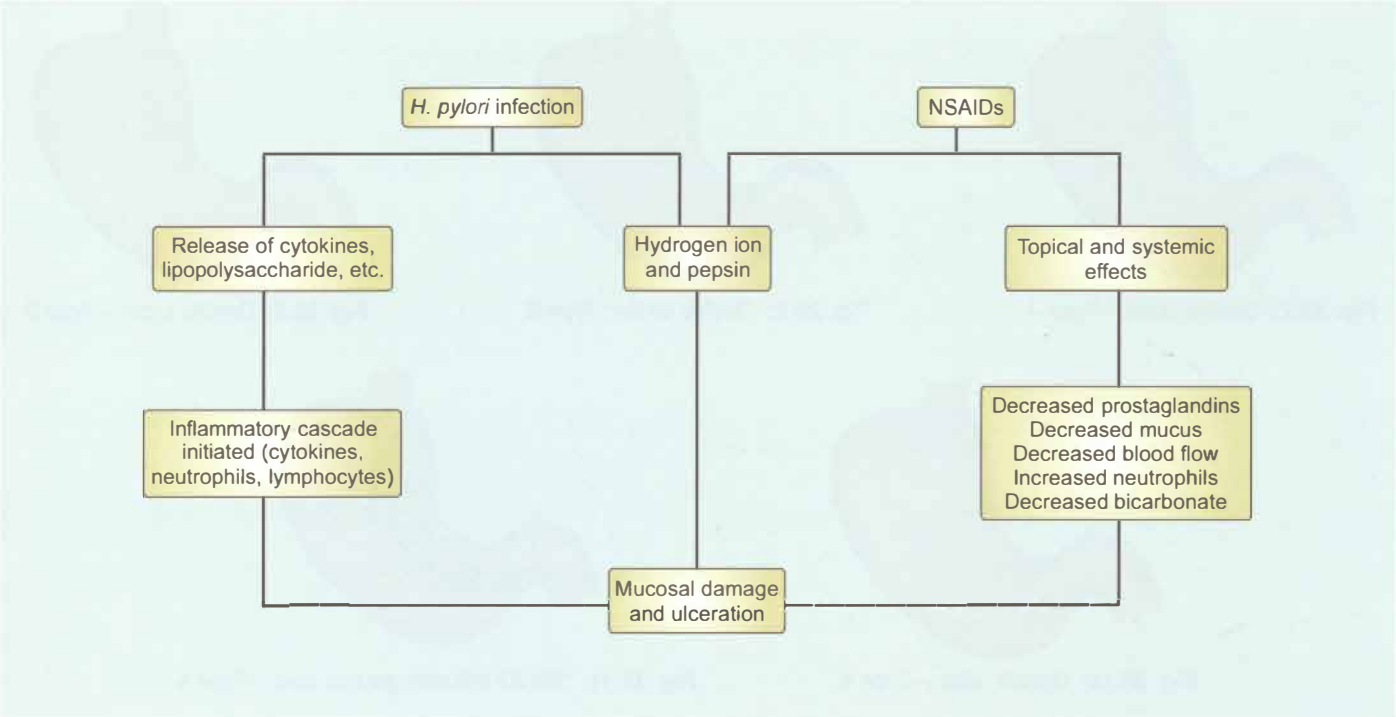


Fig. 23.6: Peptic ulceration

- Hyperparathyroidism causes increased levels of calcium which stimulates the parietal cell mass resulting in hyperacidity.

Pathophysiology of duodenal ulcer

- Duodenal ulcer is a disease of multiple aetiologies.
- The only absolute requirements are acid and pepsin secretion in combination with either infection with *H. pylori* or ingestion of NSAIDs.
- Although there is a strong correlation between parietal cell number and MAO, only patients with **duodenal ulcers have an increase in mean parietal cell number**, whereas gastric ulcer patients do not.

CHRONIC GASTRIC ULCER (Key Box 23.6)

- In majority of the patients there is no **hyperacidity**. Many patients have hypoacidity, or normoacidity.
- Ulcer occurs due to a defective gastric mucosal barrier. This barrier is a coat of thick mucus which is impermeable to pepsin.
- Prostaglandins normally present in the gastric mucosa do not allow back diffusion of hydrogen ions from the lumen.
- Nonsteroidal anti-inflammatory drugs inhibit the production of prostaglandins causing a loss of protective activity.
- This can also be damaged by smoking, spicy food, alcohol and reflux of bile into the stomach.

KEY BOX 23.6

SOME CONDITIONS THAT MAY PREDISPOSE TO GASTRIC ULCERATION

- Age older than 40, sex (female to male ratio of 2 : 1).
- Ingestion of barrier-breaking drugs such as aspirin or NSAIDs—nonsteroidal anti-inflammatory drugs
- Abnormalities in acid and pepsin secretion
- Gastric stasis through delayed gastric emptying
- Coexisting duodenal ulcer
- Duodenal gastric reflux of bile
- Gastritis, and infection with *H. pylori*
- Chronic alcohol intake, smoking
- Long-term corticosteroid therapy, infection
- Intra-arterial chemotherapy

Classification (Johnson) Types (Figs 23.7 to 23.11)

I. Acute superficial: Single or multiple (erosions)

II. Chronic (Key Box 23.7)

Type 1: Primary gastric ulcer on the lesser curvature in the antrum near the junction of oxyntic cells and central mucosa.

Type 2: Gastric ulcer with duodenal ulcer.

Type 3: Prepyloric or channel ulcer.

Type 4: High gastric ulcer (cardia, proximal stomach) < 2 cm from oesophageal junction.

Type 5: NSAID-induced gastric ulcers.

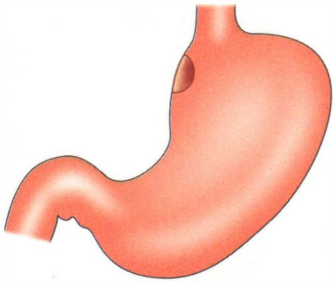


Fig. 23.7: Gastric ulcer—Type 1

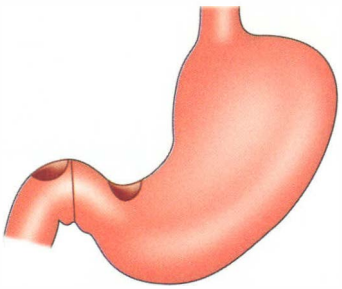


Fig. 23.8: Gastric ulcer—Type 2

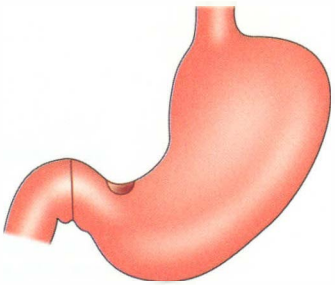


Fig. 23.9: Gastric ulcer—Type 3

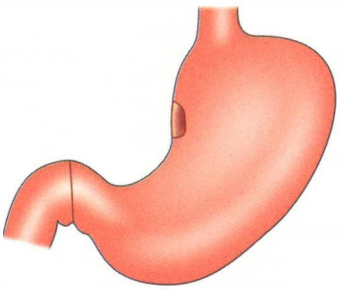


Fig. 23.10: Gastric ulcer—Type 4

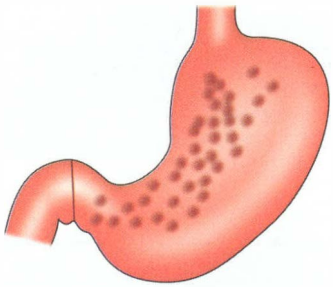


Fig. 23.11: NSAID-induced gastric ulcer—Type 5

KEY BOX 23.7

GIANT GASTRIC ULCER

- Diameter > 3 cm
- Typically lesser curvature
- Incidence of malignancy—10%
- High chances of perforation and bleeding.
- Medical treatment heals majority of cases (80%)
- Re-endoscopy biopsy is a must to look for healing and also to rule out malignancy
- Gastrectomy including ulcer bed. Add vagotomy for type 2 and type 3 gastric ulcer

PEARLS OF WISDOM

Types 2, 3 gastric ulcer are to be treated like duodenal ulcer because they are associated with hyperacidity.

Clinical features (Table 23.5)

Investigations (Figs 23.12 to 23.21)

1. **Oesophagogastroduodenoscopy (OGD):** Ulcer appears as a crater with/without slough or bleeding in their typical locations. In gastric ulcers, routine biopsy is advised to rule out malignancy. In duodenal ulcers, biopsy is done in recurrent cases to rule out *Helicobacter pylori*.

Table 23.5 Comparison of clinical features

	Chronic duodenal ulcer	Chronic gastric ulcer
1. Incidence	Common	Less common
2. Site	1st inch of 1st part of duodenum	The lesser curvature or prepyloric region
3. Pain	It is due to the acid irritating the ulcer (hunger pain). It is relieved on taking food. After 1–2 hours of food, the pain becomes severe. It is burning in nature with retrosternal radiation (heart burn) and increased salivation (water brash)	Pain occurs on taking food and it is relieved by induced vomiting. Pain is of burning nature as in duodenal ulcer
4. Vomiting	Never occurs in duodenal ulcer till the patient develops pyloric stenosis	Frequent and it occurs immediately after patient takes food
5. Weight	Weight gain	Weight loss
6. Periodicity	Common	Less
7. Haematemesis : melaena ratio	40 : 60	60 : 40
8. Incidence of malignancy	Never becomes malignant	0.5–5% (2%)
9. On examination	Tenderness in the right hypochondrium	Tenderness in the epigastrium

KEY BOX 23.8

OESOPHAGOGASTRODUODENOSCOPY(OGD)

Diagnostic (Figs 23.14 to 23.20)
- Peptic ulcers—acute and chronic
- Gastritis
- Carcinoma stomach
- Oesophageal varices, ulcers, oesophagitis
- Biopsy to rule out *H. pylori* infection
- Brush cytology and biology, yield is better

Therapeutic
- Injection of adrenaline into the bleeding vessel
- Variceal injection
- Snaring of polyps
- Electrocoagulation of bleeders
- Endoscopic cystogastrostomy
- Foreign body removal
- Percutaneous endoscopic gastrostomy (PEG)
- Endoscopic mucosal resection for early carcinoma stomatch

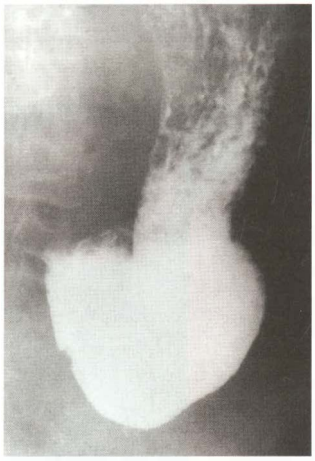


Fig. 23.13: Gastric outlet obstruction. Barium meal can detect dilated stomach

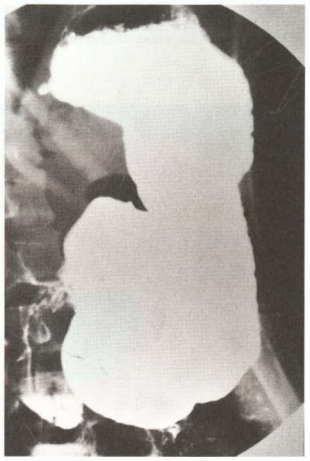


Fig. 23.14: Hugely dilated J-shaped stomach

- In long-standing duodenal ulcers, there may be narrowing of the pylorus, with stasis of food in the stomach suggestive of pyloric stenosis.
- Other uses of OGD (Key Box 23.8).

2. Barium meal study (historical—obsolete now)
- **Duodenal ulcer:** Deformed duodenal cap. **Trifoliate deformity** is seen when secondary duodenal diverticulum occurs.
- **Gastric ulcer** appears as a niche in the lesser curvature due to ulcer crater and as a notch on the greater curvature due to the spasm of stomach.
- In general benign ulcers are smoother, more regular, with rounded edges and a flat, smooth ulcer base. In malignancy, the mass protrudes into the lumen or has folds surrounding ulcer crater.
- To detect **hour glass stomach** and **gastric outlet obstruction**.

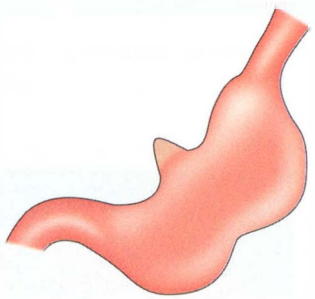


Fig. 23.12: Barium study showing niche and notch

3. Test for *Helicobacter pylori* (already discussed).

Treatment of chronic duodenal ulcer (CDU)

Aim is to decrease the pain because of its severity (by reducing acidity) and to prevent relapses (Key Box 23.9).

I. Medical line of management

1. Histamine receptor blockers (H_2 receptor blockers)
- **Ranitidine:** 150 mg twice a day, for 6 weeks. 90–95% of the healing occurs within 6 weeks. At the end of 6 weeks, 150 mg at bed time is given for a period of 3 months as maintenance therapy. Re-endoscopy can be done in between to assess the healing of the ulcer.

KEY BOX 23.9

AIMS OF THE TREATMENT
- To relieve symptoms
- To heal the ulcer
- To prevent recurrence
- To prevent complications

- **Famotidine:** 20 mg twice a day is as effective as ranitidine.
- **Roxatidine:** Can be used along with food, bronchodilators and antacids, unlike the other H_2 blockers. The dosage is 75 mg twice a day.
  - The problem with H_2 blockers is that relapse occurs if they are stopped. 80–90% healing rates occur after 6 weeks of therapy.

2. Hydrogen ion antagonists (proton pump inhibitor)
- **Omeprazole:** 20 mg once a day for 2 weeks.
- 95–99% of healing within 2–4 weeks. More rapid healing than H_2 receptor blockers.
- Patients who receive omeprazole for one month are put on a maintenance dose of ranitidine for 3 months (150 mg HS for 3 months).
- Other drugs such as esomeprazole: 40 mg/day, lansoprazole: 30 mg/day, pantoprazole: 40 mg/day are also used.
- **Pantoprazole in a dose of 40 mg/day for 4 weeks has become the first line of treatment for chronic duodenal ulcer.**

3. Regular antacids—calcium or magnesium based
- Given in high doses (120 ml/day) they will neutralise the acid (not practical). Small dose of antacids can be added to H_2 blockers. This gives psychological benefit to the patient.

4. Diet should be bland. Spicy food, coffee, alcohol, and smoking are discouraged. Discontinue aspirin or NSAIDs and other gastric irritants.

5. Eradication therapy (already discussed)

A FEW ENDOSCOPIC PICTURES

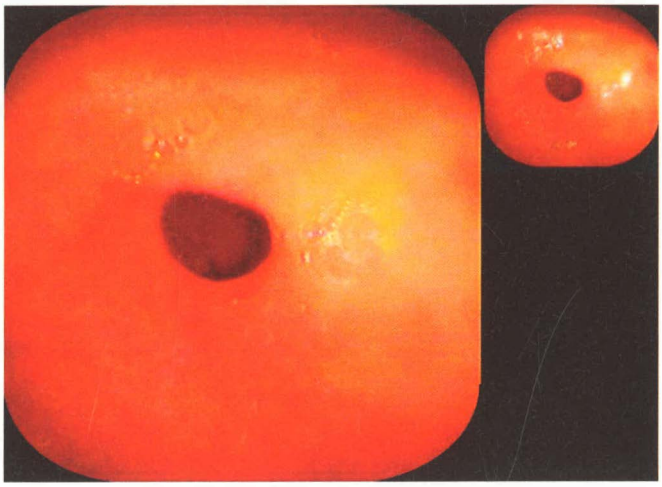


Fig. 23.15: Normal pyloric opening

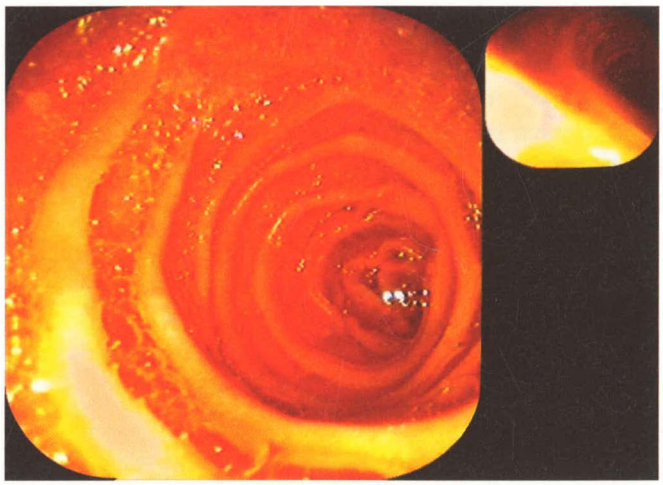


Fig. 23.16: Normal 2nd part duodenum

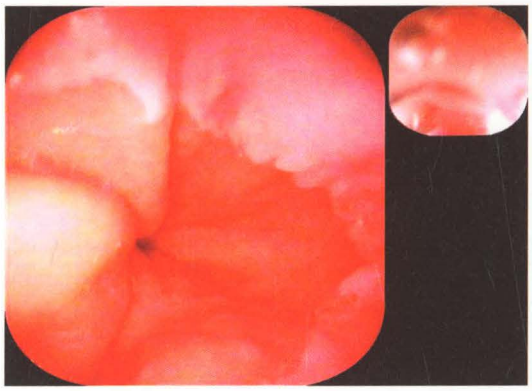


Fig. 23.17: Normal squamocolumnar junction—Z line

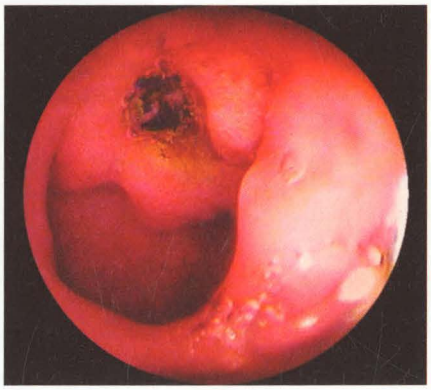


Fig. 23.18: OGD showing bleeding duodenal ulcer

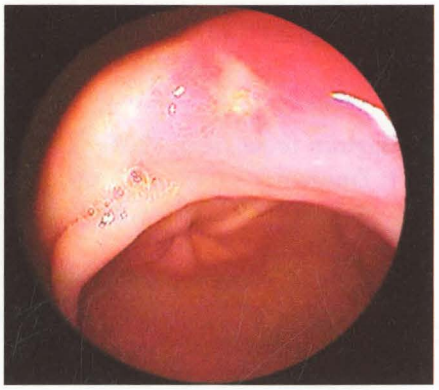


Fig. 23.19: OGD showing chronic gastric ulcer

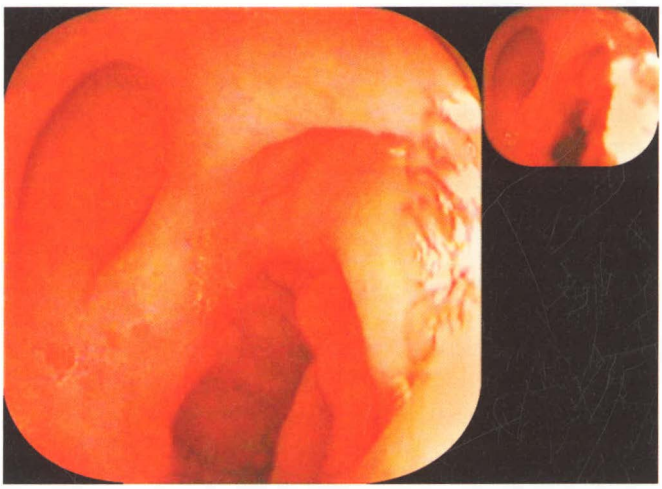


Fig. 23.20: OGD showing bleeding duodenal ulcer and diverticulum due to ulcer

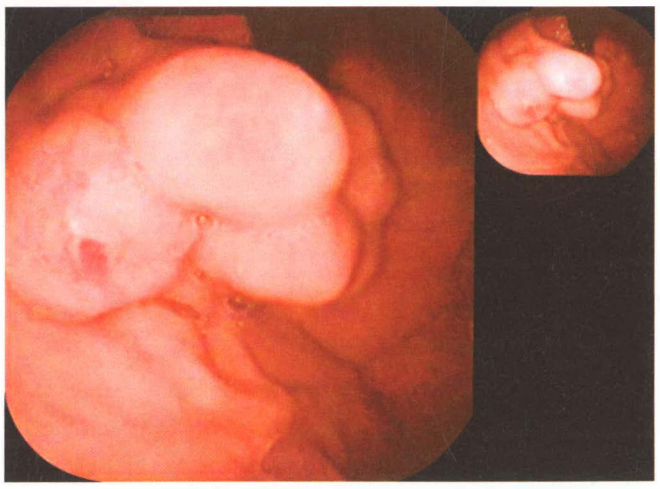


Fig. 23.21: Large gastric varices—this patient also had duodenal ulcer

(*Courtesy:* Prof Ganesh Pai, HOD, Gastroenterology, KMC, Manipal)

II. Surgical line of management

Surgery (Key Box 23.10)

1. **Highly selective vagotomy (HSV):** It is also called PCV (parietal cell vagotomy) or PGV (proximal gastric vagotomy) (Fig. 23.22).

- In this operation, **vagi are not divided at the trunks**. Both anterior and posterior vagus are identified, isolated and preserved. Their branches which run along the lesser curvature are isolated. They are anterior and posterior greater gastric nerves of Latarjet. The branches of the nerves of Latarjet supplying parietal cell mass are divided. Hence, it is called parietal cell vagotomy. The terminal fibres of the nerve of Latarjet which supply pylorus are preserved (5–7 cm of 'Crow foot').

Advantages of HSV

1. More physiological, with **minimal disturbances.**
2. No drainage procedure is required because pyloric functions are preserved.
3. Nerve supply to gall bladder and liver is not disturbed.
4. No diarrhoea as that can occur in 5–8% of cases of truncal vagotomy which can be morbid.

PEARLS OF WISDOM

It is important to note that experience of many surgeons today with HSV is 'minimum'. Hence, it is safe to do 'Vagotomy and Gastrojejunostomy (GJ)'.

Disadvantages of HSV

- This is not the procedure for prepyloric ulcer as there is a high recurrence rate.
- Complicated procedure—needs an experienced surgeon.
- Recurrence rate: 10–15%
- Rare chance of lesser curvature necrosis.

2. **Total truncal abdominal vagotomy with gastrojejunostomy of Mayo** (Fig. 23.23) **or pyloroplasty**

- This is the most popular and most commonly done operation for peptic ulcer disease. However, it is important to realise that **indications for vagotomy and GJ are becoming less and less today**.
- Transection of both vagal trunks at oesophageal hiatus denervates acid producing fundus of stomach. It also denervates the remainder of the viscera supplied by vagus including liver, biliary tree, pancreas, intestines up to mid-transverse colon.

Procedure

- **Anterior and posterior trunks of the vagus are divided** just below the diaphragm followed by a drainage procedure such as gastrojejunostomy (GJ). **Vagus is secretomotor to stomach and after vagotomy the motility of the stomach is lost and gastric stasis occurs. Hence, drainage procedure is done.**
- Posterior GJ is preferred because it gives a dependent drainage of the food contents by gravity. Classically, it is described as "Posterior, Vertical, Retrocolic, Isoperistaltic, No loop (short loop), No tension, GJ of Mayo **(PVRING)**".
- Alternatively, pyloroplasty is preferred by a few surgeons instead of GJ.
- In **Heinecke-Mickulicz pyloroplasty,** pylorus is incised longitudinally and sutured vertically. Thus, the pyloric ring becomes incompetent and wide open. Bile reflux gastritis and diarrhoea are major problems after pyloroplasty (Fig. 23.24).
- **Finney pyloroplasty** is another procedure wherein inverted U-type of incision is given involving distal stomach and duodenum and a gastroduodenal stoma is created. This type of pyloroplasty is indicated when duodenum is dilated (Fig. 23.25).

3. **Vagotomy and antrectomy** (Fig. 23.26)

- By removing the vagal stimuli and the antral gastrin, the entire stimulus to acid secretion is lost. Hence, it carries the **least recurrence rate (1%)** but carries 3–4% mortality rate. Not done routinely.

KEY BOX 23.10

INDICATIONS FOR SURGERY

1. **Intractable pain** in spite of treatment with proton pump inhibitor and not responding to anti *H. pylori* regime
2. **Frequent relapses**, *H. pylori* negative
3. **Complications** of duodenal ulcer:
   - Gastric outlet obstruction
   - Haemorrhage

Treatment of gastric ulcer

- Aim: Healing of the ulcer and relief of symptoms.
- Frequent biopsy is done to rule out malignancy.

I. Medical line of treatment can be given for duodenal ulcer, in the form of ranitidine or omeprazole. Cigarette smoking should be stopped. Drugs such as NSAID and aspirin are to be avoided. If an ulcer persists after 6 weeks, the aim is to eradicate *H. pylori* **provided malignancy is ruled out**.

II. Surgery is indicated in case of gastric ulcer persisting in spite of medical treatment.

1. **Billroth I partial gastrectomy** (Fig. 23.27)

- Partial gastrectomy is done including removal of the ulcer followed by gastroduodenal anastomosis. It has the least recurrence rate of less than 1% but mortality rate is around 1–2%.

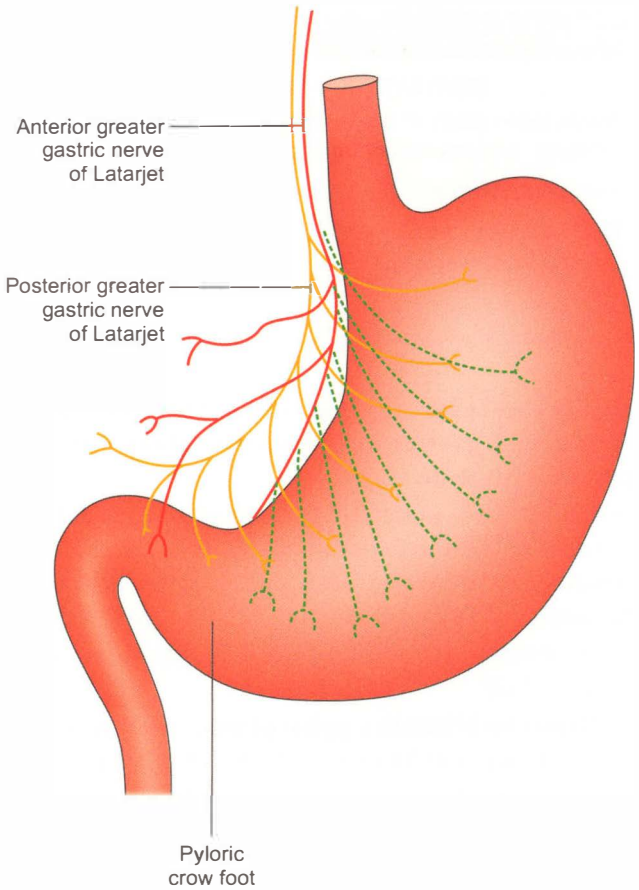


Fig. 23.22: Highly selective vagotomy—pyloric crow foot is preserved. Hence, drainage procedure is not required

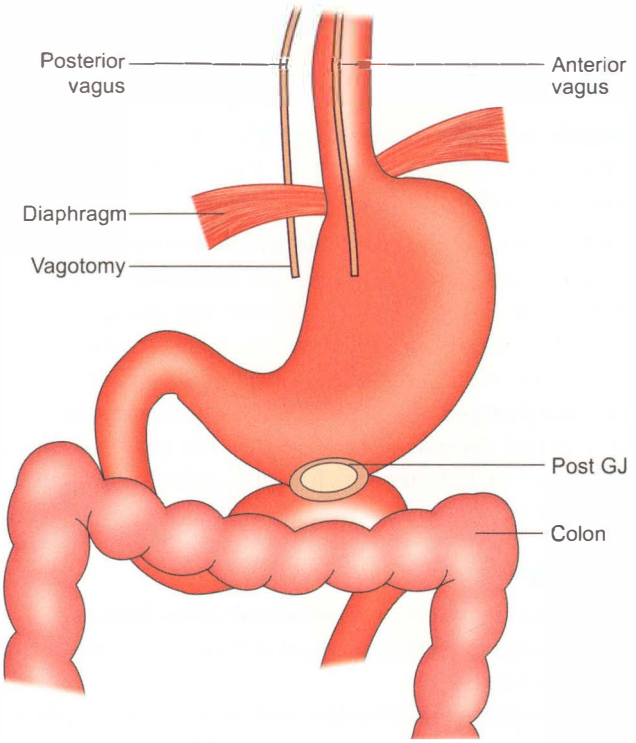
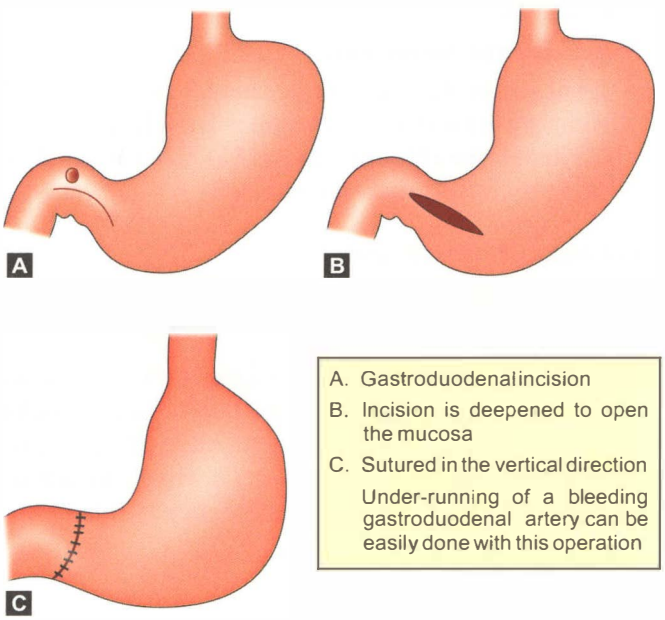


Fig. 23.23: Posterior GJ **(Mayo)**[1] with vagotomy—motility of the stomach is lost after vagotomy. Hence, drainage procedure (GJ in this picture) is required (vagus is secretomotor of the stomach)



Figs 23.24A to C: Steps of Heinecke-Mickulicz pyloroplasty

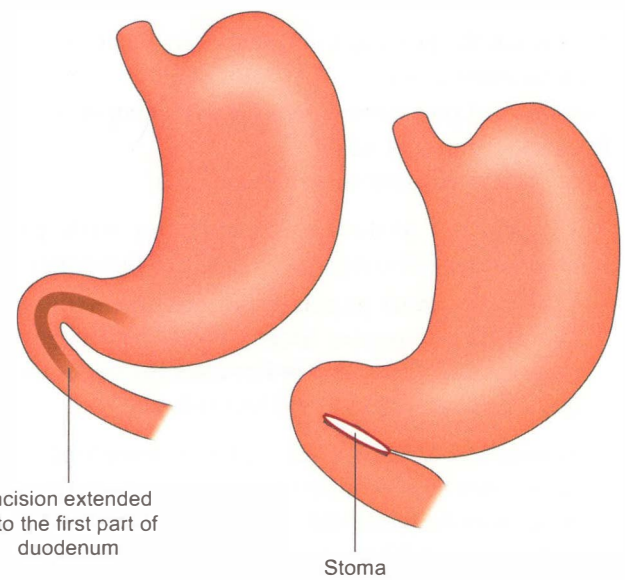


Fig. 23.25: Finney pyloroplasty

[1]About Mayo—Mayo's scissors, Mayo's forceps, Mayo's GJ, Mayo's vein, Mayo's umbilical hernia repair to be remembered.

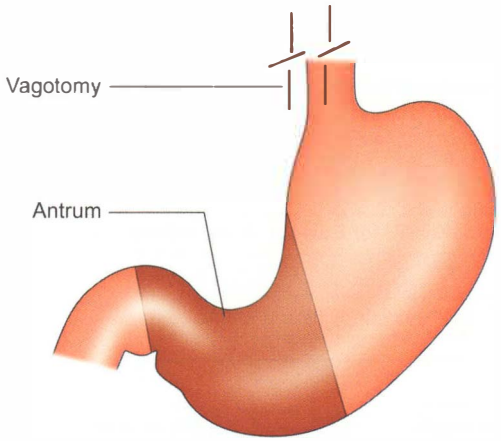


Fig. 23.26: Vagotomy with antrectomy

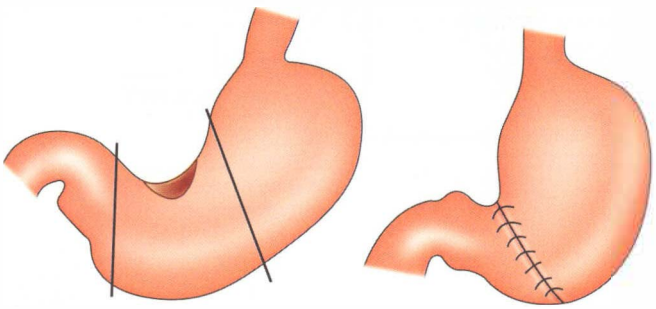


Fig. 23.27: Billroth I gastrectomy followed by gastroduodenal anastomosis

2. Billroth II gastrectomy (Fig. 23.28)

- It is indicated when the gastric ulcer is located on the lesser curvature. Here the gastrectomy is done below the ulcer and remnant of the stomach is anastomosed to a jejunal loop (gastrojejunal anastomosis). This is also described as Polya gastrectomy.

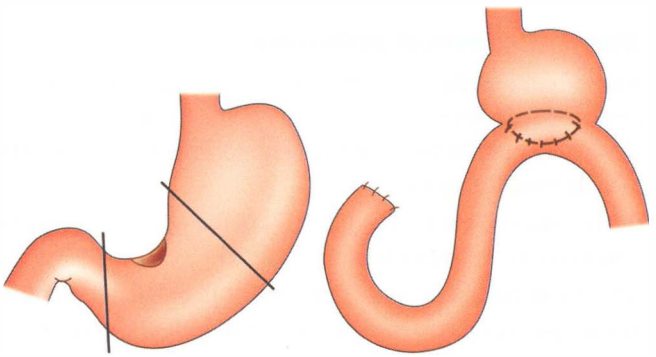


Fig. 23.28: Billroth II gastrectomy followed by gastrojejunal anastomosis

3. HSV with excision of the ulcer can be done if the experience of surgeon is good.

Type 4 gastric ulcers (*see* Fig. 23.10)

- The type 4 gastric ulcer presents a difficult management problem.
- The surgical treatment depends on the ulcer size, distance from the GE junction and the degree of surrounding inflammation.
- Whenever possible, the ulcer should be excised.
- The most aggressive approach is to perform a gastrectomy that includes a small portion of the oesophageal wall and the ulcer followed by a Roux-en-Y oesophago-gastro-jejunostomy to restore intestinal continuity.
- For type 4 gastric ulcers that are located 2 to 5 cm from the gastroesophageal junction, a distal gastrectomy with a vertical extension of the resection to include the lesser curvature with the ulcer can be performed (i.e. Pauchet procedure). After resection, bowel continuity is restored with an end-to-end gastroduodenostomy.
- **The Csendes procedure may be useful in stable patients or the Kelling-Madlener operation in unstable ones (Fig. 23.29).**
- Some have even advocated leaving the ulcer in place or locally excising it in conjunction with truncal vagotomy and pyloroplasty.

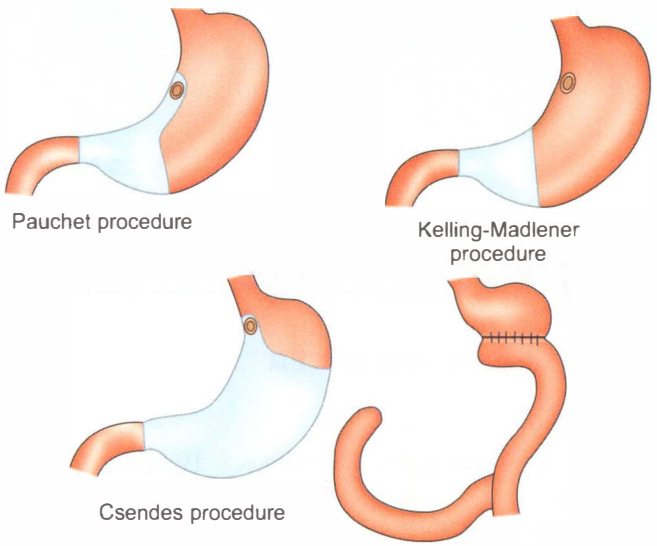


Fig. 23.29: Other types of resections for gastric ulcer

Summary of the management of peptic ulcer (Fig. 23.30)

Complications of peptic ulcer

A. Acute

- Perforation
- Haematemesis and/or melaena

B. Subacute—residual abscess

C. Chronic

- Gastric outlet obstruction (pyloric stenosis)
- Teapot deformity
- Hour glass contracture of the stomach
- Penetration into the pancreas
- Carcinoma of stomach

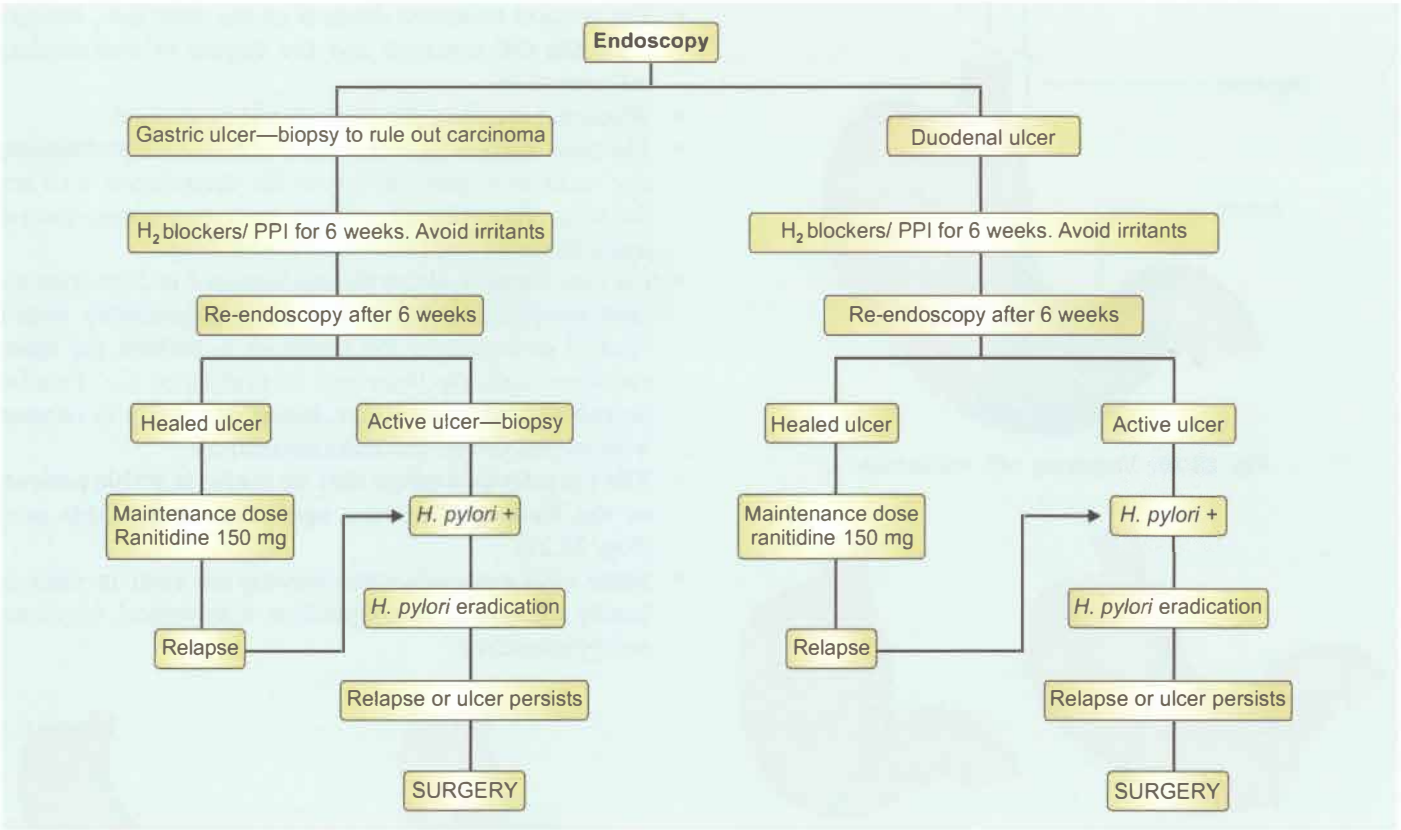


Fig. 23.30: Management of peptic ulcer

ACUTE COMPLICATIONS OF PEPTIC ULCER

PERFORATED PEPTIC ULCER

Introduction

- More common in males. The ratio is 8–10 men to 1 woman in India.
- **Anterior duodenal ulcer perforates and posterior duodenal ulcer bleeds.** An ulcer on the posterior wall of the stomach can perforate into the lesser sac.
- Usually, patients with a **long history of peptic ulcer**, suddenly complain of feeling something that has given way in their abdomen. It may be precipitated by excessive smoking alcohol, drugs, etc. Rarely a **'silent' ulcer** can also perforate (especially those patients treated with cortisone).
- Patients taking NSAIDs (often elderly) can present less dramatically.
- **Two factors** are associated with most perforated peptic ulcers: Chronic use of NSAIDs and *Helicobacter pylori* infection.
- Patients with ***H. pylori* infection** and perforated peptic ulcers tend to be **younger, with a male preponderance** and more prolonged period of dyspepsia. However, NSAIDs category of patients are elderly with equal proportion of both sexes being affected.
- Perforated peptic ulcers have a mortality rate of 5–10%.
- Perforated gastric ulcers in the elderly have 20–30% mortality rate.
- Golden time to operate is within 6 hours.

Stages of duodenal ulcer perforation

1. Stage of chemical peritonitis

- Immediately after the perforation, gastric and duodenal contents leak into the peritoneal cavity and produce severe agonising pain in the right hypochondrium. It is mainly HCl which produces pain.
- There may be an episode of coffee-ground vomitus, followed by melaena later.
- The pulse rate increases. The patient is pale and anxious.
- Blood pressure may be normal in the initial few hours.
- *Per* abdomen, there is guarding and rigidity of the abdominal wall.
- Rebound tenderness is present all over the abdomen. This sign is called **Blumberg's sign**.
- On percussion, liver dullness is obliterated because of collection of free air (gas) under the right dome of diaphragm. This stage is seen for about 2–4 hours from the time of perforation.
- Bowel sounds are usually absent.

2. Stages of reaction

- The peritoneum reacts to the chemical irritants by secreting peritoneal fluid. As a result of this, HCl and bile are diluted by the peritoneal secretions (reaction of peritoneum to the insult) resulting in an **improvement of symptoms**. Hence, it is also called **stage of delusion or stage of illusion.** This stage lasts for 3–6 hours. However, the signs are worse.
  - Pulse is feeble, more than 120/min.
  - Hypotension persists
  - Evidence of dehydration due to loss of fluid into peritoneal cavity.
  - Shifting dullness is present
  - Abdominal distension is due to fluid and paralytic ileus[1].
  - Bowel sounds are absent
  - Guarding and rigidity are worsened.

3. Stage of bacterial peritonitis

- The peritoneal contents get contaminated with gram-negative organisms resulting in bacterial peritonitis (the organisms are from the intestine itself and not from the peritoneum).
- The patient becomes severely ill, dehydrated, toxic with drawn in cheeks. The tongue is dry and coated but with bright eyes (**Hippocratic facies,** Fig. 23.31).
- Features of hypovolaemic and septicaemic shock such as feeble thready pulse, cold peripheries, shallow respiration, high grade fever and persistent hypotension are present. Gross abdominal distension, guarding, rigidity, abdominal tenderness all over suggest generalised peritonitis.

Fig. 23.31: Peritonitis—Hippocratic facies—sunken eyes, jaundice due to sepsis, nasogastric tube showing altered blood

Investigation of perforated duodenal ulcer

- Complete blood picture and electrolyte study.
- Plain X-ray chest or abdomen in **erect position** shows collection of free gas under the right dome of diaphragm, in majority of cases. If patient is unable to stand, left lateral decubitus films are taken (Figs 23.32 and 23.33).
- When in doubt, request a CT scan with contrast which can demonstrate pneumoperitoneum, fluid in the abdomen, site of perforation and some surprises also.

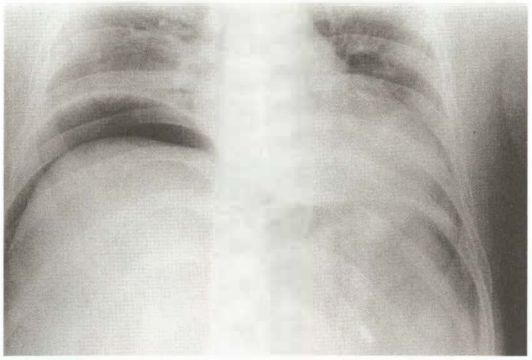


Fig. 23.32: Plain X-ray abdomen erect showing collection of free gas under the right dome of the diaphragm

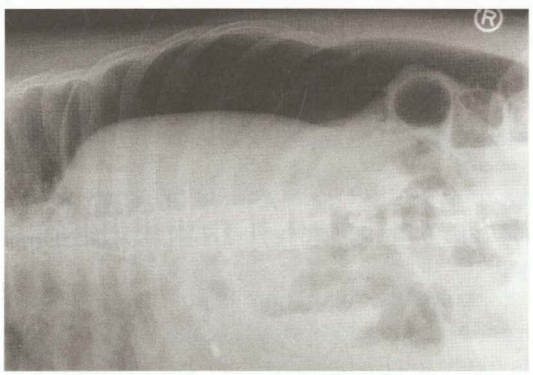


Fig. 23.33: Plain X-ray abdomen lateral decubitus showing collection of free gas under the abdominal wall

Treatment (ABCDEF)[2]

A **A**spiration of stomach contents with Ryle's tube to reduce further contamination and to decrease biliary and pancreatic juice.

B **B**lood grouping and cross-matching may be necessary for surgery.

C **C**harts: Temperature, pulse, BP, respiration, urinary output (urinary bladder is catheterised using a Foley's catheter).

D **D**rugs:
- Injection ampicillin 500 mg IV, stat and 6th hourly against gram-positive organisms.

[1]It resembles a strike by employees paralysing the work of a factory in response to an insult.

[2]Students should remember that ABCDEF are the basic principles of treatment of any acute abdomen. In majority of cases of acute abdomen, these principles can be applied with minor modifications.

- Injection gentamicin 60–80 mg IV, 8th hourly against gram-negative organisms.
- Injection metronidazole 500 mg IV, 8th hourly to treat anaerobic organisms.
- Cephalosporins can also be used depending upon the severity of the shock.

E Exploratory laparotomy is done through a midline incision. The perforation is identified and closed with interrupted nonabsorbable silk sutures, which is strengthened by **placement of omentum** (Fig. 23.34). Peritoneal toilet/wash is given to avoid residual abscess. Abdomen is closed with a drain which is removed after 3–5 days. If it is a large gastric ulcer, it is better to do a gastrectomy, if condition of the patient permits.

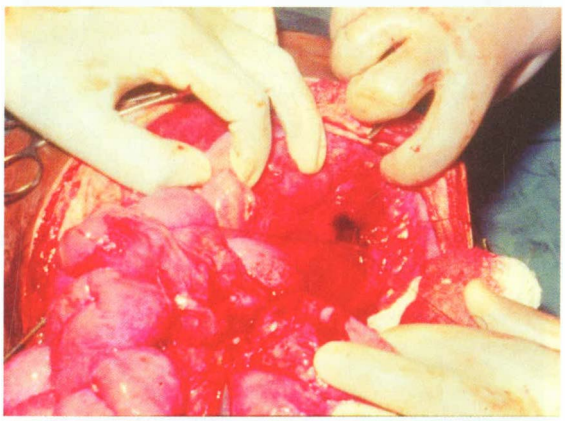


Fig. 23.34: Duodenal ulcer perforation—closure

- Vagotomy and GJ is not done at this stage as the general condition of the patient is very poor and there is peritoneal sepsis.
- Postoperatively, the patient is put on anti-ulcer drugs.
- An endoscopy is done after 2 months. If the ulcer persists, it is likely to be a chronic ulcer and an elective operation such as vagotomy and GJ is done. Simple suturing cures majority of acute ulcers.

F Fluids are given preoperatively to treat dehydration and postoperatively for 3–4 days till the paralytic ileus settles down (soft abdomen and bowel sounds present).

Early cases of perforation

It can also be managed by laparoscopic closure of perforation with peritoneal drainage. In fact, a thorough wash is really possible with a laparoscope.

Subacute

Perforation with abscess. Some patients present late to the hospital with features of sealed perforation.

HAEMORRHAGE FROM PEPTIC ULCER

- Haemorrhage from peptic ulcer can be chronic which causes anaemia, or acute, resulting in massive haematemesis and melaena.
- It is the **posterior duodenal ulcer which commonly bleeds**, because it erodes into the gastroduodenal artery which runs posterior to the duodenum. A gastric ulcer on the lesser curvature erodes into one of the branches of left or right gastric artery.

Precipitating factors for haemorrhage

1. **Chronicity**, results in destruction of the layers of the stomach, exposing the vessel.
2. Sudden, severe acid peptic digestion brought about by **irritants** such as alcohol, drugs, etc.
3. **Atherosclerosis:** Sclerotic artery does not contract, resulting in massive haemorrhage.

Characteristics of individuals at an increased risk of developing acute GI bleeding

- Increased age
- Male sex
- Cardiovascular disease
- Diabetes mellitus and renal disease
- Increased number of medications
- Oral anticoagulant use

Clinical features of bleeding peptic ulcer

1. Previous history of abdominal pain of peptic ulcer disease.
2. History of haematemesis or melaena (black tarry stools), one or more attacks.
3. There may be features of haemorrhagic shock such as feeble, thready pulse, hypotension, syncope.
   - Oliguria, due to inadequate renal perfusion.
   - Brainstem hypoxia results in change in rate and depth of respiration.
   - There may not be any abdominal signs. However, due to accumulation of blood in the intestines and stomach, mild distension may be present. **Perforation produces abdominal signs and haemorrhage produces systemic signs.**

BLEAD risk classification

The following are some of the factors associated with increased morbidity and mortality (Key Box 23.11).

KEY BOX 23.11

BLEAD RISK CLASSIFICATION

- **BL**ood pressure low
- **E**levated prothrombin time
- **A**ltered mental status
- **D**ysfunction—myocardial, renal, comorbid disease.

Forrest classification of endoscopic appearance of bleeding ulcers

Type	Description
Ia	Spurting, bleeding
Ib	Nonspurting, active bleeding
IIa	Visible vessel
IIb	Nonbleeding ulcer with overlying clot
IIc	Ulcer with haematin—covered (black) base
III	Clean ulcer base

As previously mentioned, endoscopy provides the opportunity not only for diagnosis but also for therapy.

Management

- Emergency upper OGD is done to confirm the diagnosis. If the source cannot be detected due to large clots or massive bleeding, it can be repeated a few hours after a stomach wash and blood transfusion.
- Emergency upper OGD should be done within 12–24 hours of bleeding depending upon condition of the patient.
- Resuscitation is more important than an urgent endoscopy.
- Since elderly patients cannot tolerate shock well, decision to control bleeding surgically must be taken early.

I. Conservative line of management

1. Emergency replacement of blood, after initial resuscitation with a plasma expander.
2. Ryle's tube is passed and cold saline stomach wash is given to produce vasoconstriction.
3. Cold antacids are given every 2nd hourly, about 10–20 ml.
4. IV ranitidine 50 mg, 8th hourly or IV pantoprazole 40 mg is given to reduce acidity.
   - Majority of cases respond to conservative line of management within 48 hours.

II. Nonsurgical treatment

1. Laser coagulation

- It can arrest the bleeding without direct tissue contact. Nd:YAG laser has been used more commonly because it can penetrate tissue more deeply compared to argon laser which penetrates very superficial tissues. The success rate of laser coagulation is around 80%.

2. Sclerotherapy (Key Boxes 23.12 and 23.13)

- Epinephrine (1:10,000) arrests bleeding by vasoconstriction.
- 2% ethanolamine, a sclerosant causes dehydration and shrinkage of surrounding tissues.
- It also produces inflammation and thrombosis of the bleeding vessel (Fig. 23.35).
- This is the most popular method. The success rate is around 80–90%. It is a cheap and easy treatment.

3. Haemoclip application (Fig. 23.36)

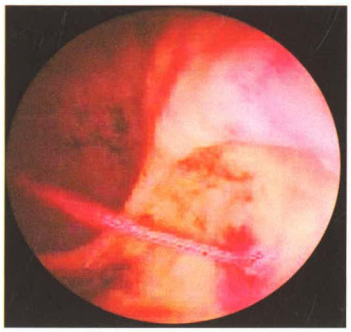


Fig. 23.35: Bleeding vessel

Fig. 23.36: Haemoclip applied

4. Bipolar electrocoagulation—failure rate is 50%
   - Surgical eradication of *H. pylori* prevents rebleeding.

III. Surgical control of bleeding peptic ulcer (Key Boxes 23.13 and 23.14)

Indications

1. Failure of endoscopic haemostasis—prognostic factors are given below.
2. Rebleeding in the hospital (rebleeding is more common in gastric ulcer patients).
3. Bleeding requiring transfusion of more than 2000 ml blood in 24 hours (6 units).
4. Elderly patients with rebleeding.
5. Massive haemorrhage leading to shock or cardiovascular instability.
6. Recurrent haemorrhage requiring hospitalisation.

KEY BOX 23.12

ENDOTHERAPY

- Bipolar electrocoagulation—failure rate—50%
- Inj. sclerotherapy—failure rate—20%
- Haemoclip application

KEY BOX 23.13

ENDOSCOPIC PROGNOSTIC FACTORS

- Visible level: 40–60% rebleeding ulcer > 2 cm
- Adherent clot: 20% rebleeding
- Flat pigmented spot: 10% rebleeding
- Clean ulcer base: Rarely bleeding

KEY BOX 23.14

ROLE OF SURGERY

- Explore when endotherapy fails
- Explore when rebleeding occurs in the elderly
- Explore an unstable patient
- Gastroduodenotomy, under-running of ulcer base for CDU and partial gastrectomy for gastric ulcer.

Types of surgery

1. Surgery for bleeding duodenal ulcer

- Laparotomy and **anterior gastroduodenotomy.**
- Visualise the bleeding ulcer in the first part of duodenum
- **Under-running** of the ulcer base by direct suture or 4 quadrant ligation of gastroduodenal artery by using nonabsorbable sutures (Fig. 23.37).
- **Gastroduodenotomy incision is converted into a pyloroplasty** followed by vagotomy which completes the treatment.

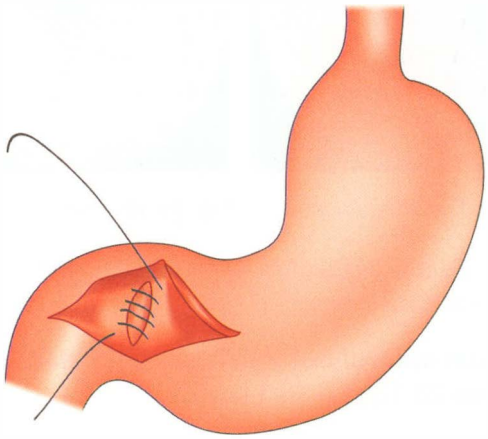


Fig. 23.37: Under-running of the duodenal ulcer

2. Surgery for bleeding gastric ulcer (benign)

- Laparotomy, gastrotomy and visualise the bleeding ulcer.
- Under-running of the ulcer base. There are chances of rebleeding with this method.
- **Partial gastrectomy** is the best treatment provided general condition of the patient is good (Figs 23.38 and 23.39). Otherwise, local excision of the ulcer, vagotomy followed by GJ or pyloroplasty can also be done.

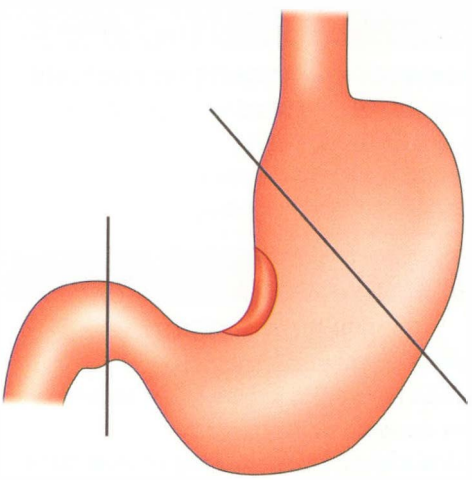


Fig. 23.38: Partial gastrectomy for gastric ulcer

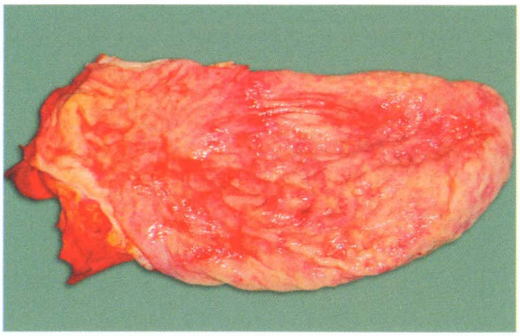


Fig. 23.39: Partial gastrectomy for bleeding gastric ulcer

- Haemostatic methods currently employed include thermotherapy (heater probe, multipolar or bipolar electrocoagulation) as well as injection of ethanol or epinephrine solutions.
- When the bleeding is controlled, long-term medical therapy includes antisecretory agents, usually in the form of a proton pump inhibitor, in addition to testing for *H. pylori* with treatment if positive.
- If *H. pylori* is present, documentation of eradication should be performed after therapy.

DIFFERENTIAL DIAGNOSIS OF HAEMATEMESIS (UPPER GI TRACT BLEEDING)

- 80% bleeding is from upper GI tract.
- Almost 20% is from lower GI tract (colon)
- 1% from small intestines.
- Bleeding proximal to ligament of Treitz.
- Haematemesis refers to vomiting of fresh red blood. Melaenemesis refers to vomiting of dark altered blood. However, both are included under upper GI tract bleeding. Small bowel, even though is a midgut structure, some of its lesions can produce haematemesis. Hence, they are also included under this heading.

Causes (Fig. 23.40)

1. Oesophageal causes

- Reflux oesophagitis
- Mallory-Weiss syndrome
- Oesophageal varices
- Cancer of oesophagus, leiomyoma oesophagus

2. Gastric causes

- Gastric ulcer
- Gastric varices
- Acute erosive gastritis
- Gastric cancer
- Stromal tumours—**GIST**
- Lymphoma
- Arteriovenous malformation

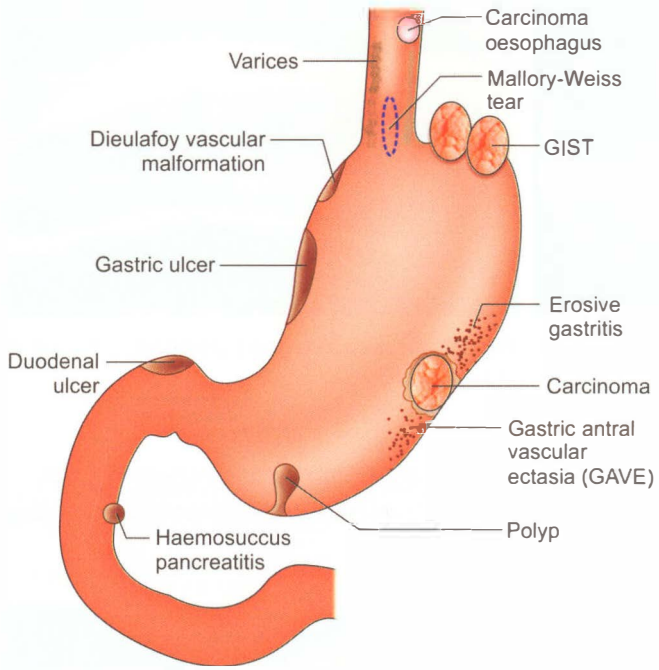


Fig. 23.40: Various causes of haematemesis

- Gastric polyp
- Dieulafoy vascular malformation
- Gastric antral vascular ectasia (GAVE)

3. Duodenal causes

- Duodenal ulcer
- Arteriovenous malformation
- Duodenal carcinoma
- Aortoduodenal fistula diverticulae
- Polyps

4. Other rare causes

- Purpura
- Haemophilia
- Haemobilia
- Pseudoaneurysms due to acute pancreatitis.

PEARLS OF WISDOM

Acute erosive gastritis, chronic peptic ulcer and oesophageal varices constitute almost 90% of the cases.

Initial assessment (Table 23.6)

Investigations (Fig. 23.41)

- **Fibreoptic endoscopy** should be done on an emergency basis within 6 to 36 hours of admission. It can diagnose variceal bleeding, erosive gastritis, bleeding peptic ulcer, carcinoma stomach, etc.
- Only when endoscopy cannot yield any diagnosis, selective coeliac angiography should be done which can detect uncommon causes such as angiodysplasia of the stomach or bleeding from a rolling hernia, etc. Barium study is done to rule out intestinal causes in less urgent cases.
- **Isotope studies:** Intravenous injection of **^{99m}Tc pertechnetate** can demonstrate hypertrophic gastric like mucosa in **Meckel's diverticulum**.
- **CT angiogram is the gold standard** investigation when haemobilia or haemosuccus pancreatitis is suspected (Figs 23.42 to 23.44).

Treatment

Initial management is to treat the shock in the line discussed for peptic ulcer haemorrhage.

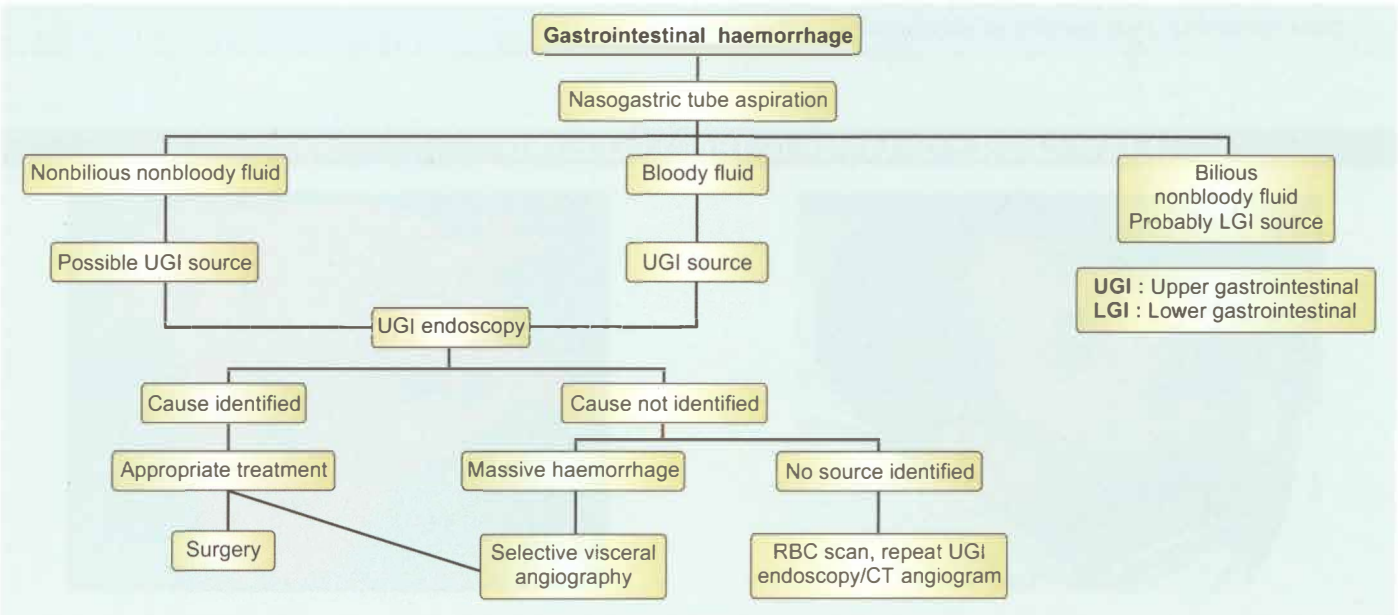
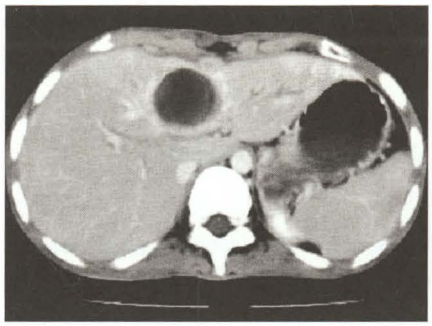
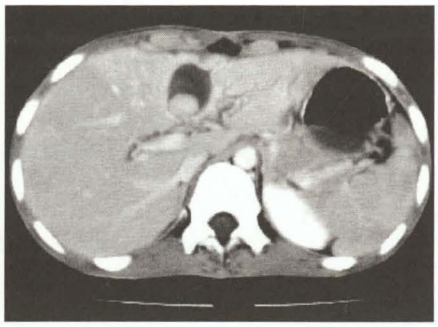


Fig. 23.41: Investigation algorithm of UGI bleeding (*Courtesy:* Dr Sibacis Bisoi, Asst Professor, KMC, Manipal)





Figs 23.42 and 23.43: CT hepatic angiogram demonstrating extravasation of dye into a pseudoaneurysm of a branch of middle hepatic artery and thrombus—case of haemobilia

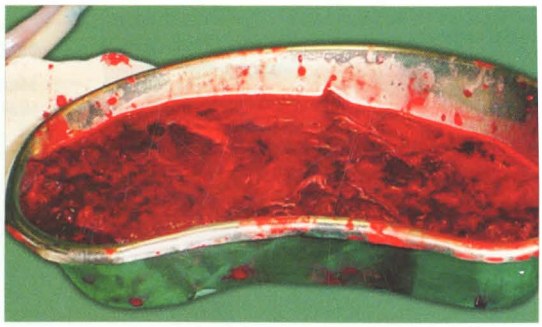


Fig. 23.44: Massive haematemesis in this patient of haemobilia

Table 23.6 Initial assessment

History	Probable causes
1. Abuse of drugs and alcohol	Acute erosive gastritis
Previous abdominal pain	Chronic peptic ulcer
H/o jaundice, liver cell failure	Oesophageal varices
Anaemia, loss of weight, loss of appetite	Carcinoma stomach
Violent vomiting—haematemesis	Mallory-Weiss syndrome
2. General physical examination	
• Testicular atrophy, gynaecomastia	• Cirrhosis of the liver
• Palpable left supraclavicular node	• Carcinoma stomach
• Arthritis—multiple joint involvement	• Acute erosive gastritis (use of NSAID)
• Purpuric spots (Fig. 23.45)	• Bleeding tendencies
3. Abdominal examination	
• Palpable spleen, ascites	• Portal hypertension
• Palpable stomach mass	• Carcinoma stomach
• Tenderness in the epigastrium	• Peptic ulcer disease
4. Assessment of haemorrhage	
Massive haemorrhage (more than 1,000 ml of blood)	Portal hypertension, acute erosive gastritis, chronic peptic ulcer
Moderate haemorrhage (500–1000 ml of blood)	Chronic peptic ulcer
Mild haemorrhage (less than 500 ml of blood)	Other causes

Fig. 23.45: Purpuric spots

A FEW CAUSES OF UPPER GI TRACT BLEEDING (Figs 23.46 to 23.51)

Fig. 23.46: Chronic duodenal ulcer with massive UGI bleeding. Diagnostic use of nasogastric tube is to detect active bleeding

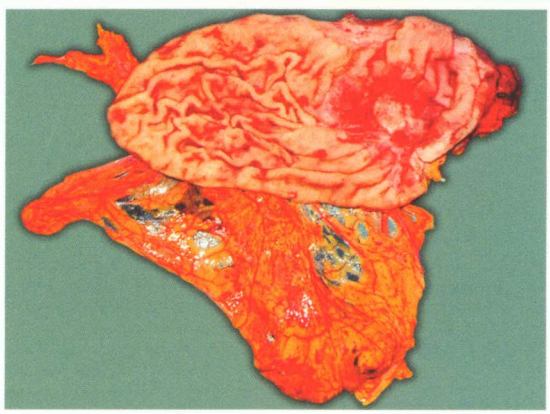


Fig. 23.47: Subtotal gastrectomy specimen showing bleeding gastric ulcer

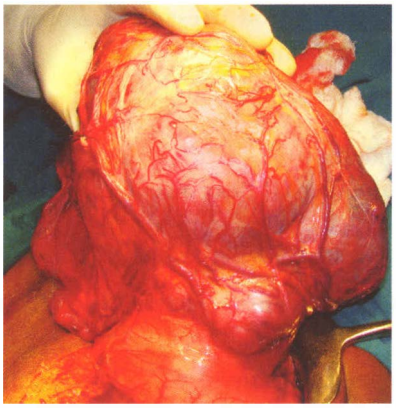


Fig. 23.48: Upper GI bleeding due to GIST of stomach

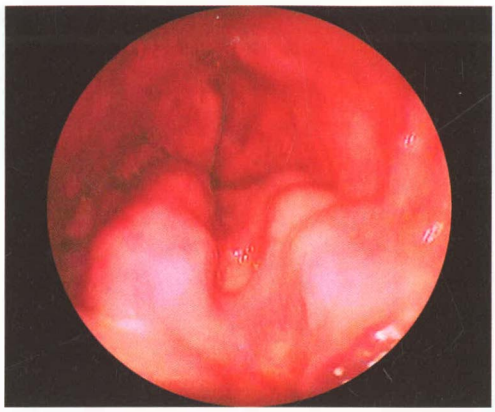


Fig. 23.49: Grade III oesophageal varices with bleeding

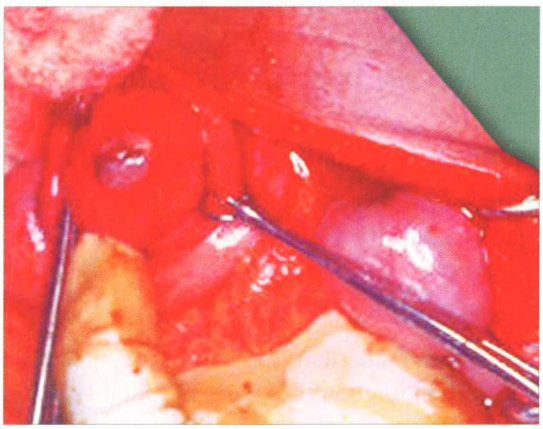


Fig. 23.50: Dieulafoy vascular malformation

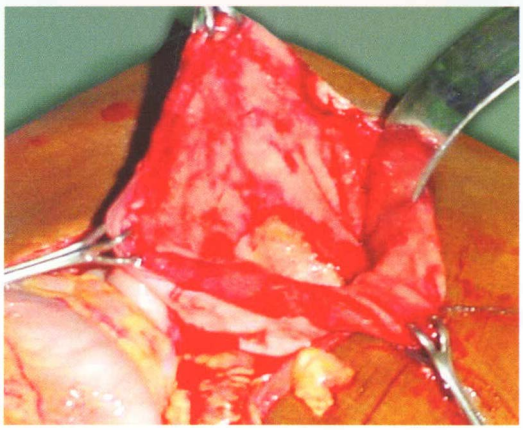


Fig. 23.51: Giant gastric ulcer with bleeding

Indications for surgery

1. Elderly patients with rebleed in the hospital
2. Rarity of blood groups
3. Spurting vessel in an endoscopy

Surgery

- Surgical management of individual case has been discussed along with the concerned chapter. However, summary of the treatment is discussed in Table 23.7.
- Thus, upper GI bleeding can occur due to various causes. However, acute erosive gastritis, chronic peptic ulcer and oesophageal varices are the three important causes of bleeding. Endoscopy is the investigation for the diagnosis of upper GI bleeding (Fig. 23.52). Today, most of the upper GI bleeding is managed either in the form of injection sclerotherapy, laser coagulation, or with powerful H_2 blockers or proton pump inhibitors. In appropriate cases, surgery is definitely indicated. Some common conditions which give rise to haematemesis are discussed in the following pages.

Table 23.7 Surgery for haematemesis

Causes	Conservative → Failure →	Surgical method
1. Acute erosive gastritis	Yes	Rarely—gastrectomy
2. Chronic duodenal ulcer	Yes	Vagotomy, under-running, pyloroplasty
3. Chronic gastric ulcer	Yes	Partial gastrectomy
4. Mallory-Weiss syndrome	Yes	Suturing of the tear
5. Cancer of the stomach	No	Gastrectomy
6. Duodenal polyp	Endoscopic snaring	Surgery, if endoscopic facility not available
7. Haemobilia	Yes, therapeutic embolisation	Ligation of feeding vessel
8. Variceal bleeding	Yes, sclerotherapy	Devascularisation
9. Haemosuccus pancreatitis	Yes, therapeutic embolisation	Ligation of pseudoaneurysm
10. Dieulafoy lesion	Yes, injection sclerotherapy	Wide excision

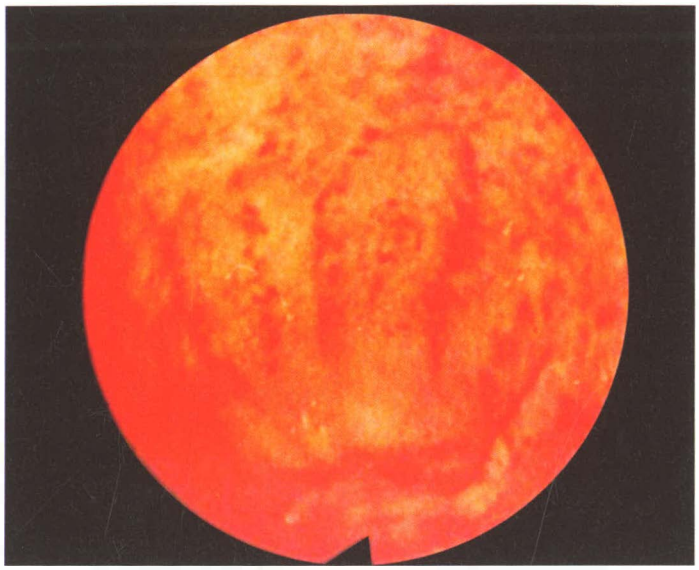


Fig. 23.52: Endoscopy showing erosions

ACUTE PEPTIC ULCER

They are also called acute erosive gastritis/acute stress ulcers/ acute steroidal ulcers and **Acute Gastric Mucosal Lesions (AGML)**.

Aetiology

- Drugs—aspirin, analgin, steroids, phenylbutazone.
- Any stress or acute infection which results in sepsis.
- Following tetanus
- After burns, they are called **'Curling's ulcer'.**
- After head injury or neurosurgical operations, they are called **'Cushing's ulcer'.**
- Following **hypotension** and shock, there is **hypoperfusion,** which results in **mucosal ischaemia** and acute stress ulcers. Example: Myocardial infarction, trauma, sepsis.
- Excessive consumption of spirits and smoking.
- Respiratory failure can also produce acute erosions.

Pathology

There is a diffuse mucosal damage and disruption of gastric mucosal barrier. Reflux of bile also may be a precipitating factor. This results in acute erosions when they are 1 to 2 mm or acute ulcers when they are 1 to 2 cm in size, shallow and well-demarcated. The entire stomach is involved by these ulcers.

Clinical features

- Dyspepsia due to minor bleeding.
- Haematemesis—sometimes massive, fresh bleeding can produce hypotension and shock.
- Acute abdominal pain due to acute erosions or perforation of an acute ulcer.
- In ICU: **Drop in blood pressure,** nasogastric aspirate o blood and/or melaena.

Diagnosis

Emergency endoscopy to confirm diagnosis and to rule ou other causes of upper GI tract bleeding.

Treatment of acute peptic ulcers with bleeding

1. **Admission in intensive care unit (ICU)** (Key Box 23.15).
2. **Ryle's tube** to be introduced first and empty the stomach. It helps in reducing vomiting and sometimes aspiration of gastric contents into the lungs also. Ice cold water or saline stomach wash (vasoconstriction) reduces bleeding.
3. **Regular antacids** 20–30 ml, 2nd hourly, preferably cold antacids.
4. **Replacement of blood** specifically coagulation factors.
5. **Injection ranitidine** 50 mg IV, 8th hourly or **pantoprazole 20 mg IV**. Majority of cases respond to conservative line of treatment.
6. **Endotherapy:** If there is solitary bleeding site.
7. **Surgery:** With modern investigations and their therapeutic role in addition to good ICU care, availability of blood and nursing care, almost all cases of erosive gastritis can be managed without surgery. In spite of above treatment if the condition deteriorates, a total gastrectomy or a subtotal gastrectomy may be necessary (very, very rare) as a life-saving measure.

KEY BOX 23.15

VERY IMPORTANT MEASURES IN ICU

- Correct coagulopathy
- Improve oxygenation
- Blood transfusion
- Control sepsis

MALLORY-WEISS SYNDROME

Aetiopathogenesis (Key Box 23.16)

- It occurs due to a tear in the gastric mucosa near the oesophagogastric (OG) junction.
- Also called partial thickness mucosal rupture.
- In Boerhaave syndrome, all layers are involved in a tear.

Clinical features (Fig. 23.53)

- The patient is usually a middle-aged male who, after consumption of alcohol, vomits the food contents first. During the course of vomiting because of straining and

KEY BOX 23.16

CONDITIONS WHEREIN MALLORY-WEISS TEAR IS SEEN

- **S**pirit or alcohol
- **P**ancreatitis
- **I**nfarction—myocardial
- **R**enal failure
- **I**nfection—cholecystitis
- **T**umour—pregnancy

Remember as **SPIRIT**

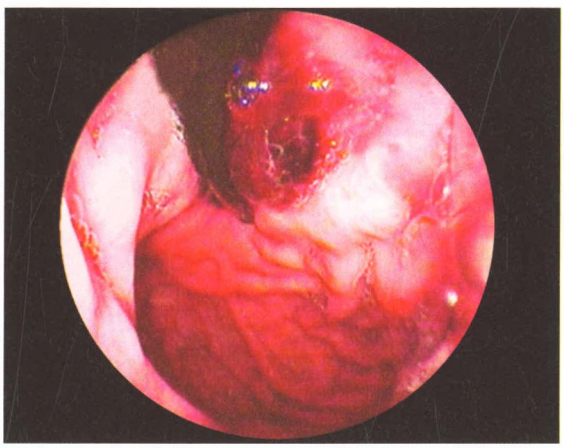


Fig. 23.53: Mallory-Weiss tear—with the endotherapy bleeding stopped (*Courtesy:* Dr Filipe Alvares, Gastroenterologist, KMC, Manipal)

retching, a tear develops near the oesophagocardiac junction. Hence, the **second vomitus contains blood**.

- Sometimes, the bleeding can be so massive to produce hypotension and shock. In 90% of cases, the bleeding stops spontaneously.

Treatment

- Urgent resuscitation of haemorrhagic shock.
- Endoscopy to confirm the diagnosis—tear is seen in the lesser curvature.
- Ryle's tube aspiration for 48–72 hours.
- Endotherapy—injection adrenaline 1:10,000 dilution is effective. If not, haemoclipping can be done.
- Multipolar electric coagulation
- When all other measures fail, **high gastrotomy** and underrunning is necessary with 2–O silk sutures.

DIEULAFOY VASCULAR MALFORMATION

- In this condition, an unusually large artery runs in the submucosa which lies in close contact with mucous membrane (Fig. 23.54).
- Mucosal erosion precipitates the bleeding.
- In more than 80% cases, bleeding occurs within 6 to 8 cm from OG junction, at the lesser curvature.
- Endoscopy and endotherapy should be tried and if necessary, repeated also.
- Sudden, massive, painless, recurrent haematemesis with hypotension is the presenting feature.
- Angiography to confirm the bleeding lesion and gelfoam embolisation must be tried first.
- Failure to achieve control of the bleeding—**gastrotomy and wide excision** of the lesion should be done.

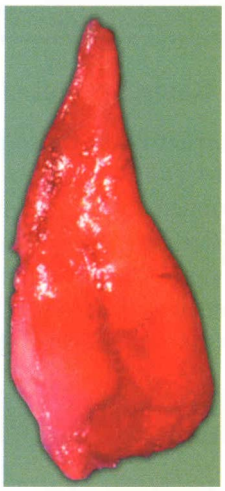


Fig. 23.54: Dieulafoy vascular malformation—excised

GASTRIC ANTRAL VASCULAR ECTASIA (GAVE)

- It is an uncommon cause of chronic GI bleeding resulting in iron deficiency anaemia.
- It is also called **watermelon stomach** because of the presence of long red streaky area which are present in the stomach resembling a watermelon (Key Box 23.17).

Subacute complication of peptic ulcer

A small perforation of peptic ulcer which is sealed off by omentum may result in a residual abscess in one of the subphrenic spaces. It responds to conservative treatment. Otherwise, percutaneous drainage can be done with ultrasonographic guidance.

KEY BOX 23.17

WATERMELON STOMACH (GASTRIC ANTRAL VASCULAR ECTASIA)

- **W**omen in their 50s are commonly affected
- **A**ntrum is commonly involved
- **T**IPSS (page 542) in patients with portal hypertension
- **E**ctasia of antral vessels gives rise to UGI bleeding
- **R**ed parallel stripes on the mucosal fold are characteristic
- **M**ucosal fibromuscular hyperplasia and hyalinisation are present
- **E**ndoscopy is the investigation of choice
- **L**iver disease in 25% patients—cirrhotic men
- **O**ther diseases—autoimmune connective tissue disorder may be associated with this—also *Helicobacter pylori*
- **N**o control of bleeding—antrectomy may be required

Remember as **WATERMELON**

CHRONIC COMPLICATIONS OF PEPTIC ULCER

I. Gastric outlet obstruction

Earlier it was called pyloric stenosis. However, gastric outlet obstruction is a better word. Chronic cicatrisation of a duodenal ulcer or juxtapyloric ulcer results in narrowing of pyloric antrum which is described as pyloric stenosis. In India, pyloric stenosis is more common in South Indian patients, who usually present with a long history of duodenal ulcer and a recent history of vomiting (Key Box 23.18).

KEY BOX 23.18

PYLORIC STENOSIS

- Pyloric stenosis in CDU—**Misnomer**
- Stenosis is very often found in the **first part of duodenum**
- In cases of pyloric channel ulcer, true pyloric stenosis occurs
- Metabolic changes, such as paradoxical aciduria (*see* below) are usually seen in patients with ulcer, **not in carcinoma** because of relative achlorhydria in the latter.

Symptoms

- Classical **hunger pain** of duodenal ulcer disappears. It may be replaced by a dull aching pain because of gastric distension. Colicky pain is due to hyperperistalsis of stomach.
- **Vomiting** is profuse, projectile, persistent, foul-smelling (because of stasis) and nonbilious.
- There may be **distension of upper abdomen** with epigastric fullness.

Signs

1. **Visible gastric peristalsis (VGP): Stomach that you see**
   - It is a wave of contraction of the stomach which starts in the left hypochondrium, runs across the umbilicus and ends in the right hypochondrium. These contractions can be felt—**stomach that you feel**. Presence of VGP is diagnostic of pyloric stenosis (right to left peristalsis is seen in left-sided obstructive colonic tumours). **If VGP is not seen, it can be made prominent by:**
   - Asking the patient to drink at least 500–1000 ml of water (It is difficult. Try and see!).
   - Stimulating the abdomen by flicking movement.
2. **Succussion splash**
   - Should be done on 'fasting' stomach. This test should be done before asking the patient to drink water.
   - In pyloric stenosis there is always residual fluid in the stomach, which gives a splashing sound that can be heard with/without stethoscope—**stomach that you hear.**

PEARLS OF WISDOM

Thus, the stomach which is seen, felt and heard is diagnostic of pyloric stenosis.

3. **Auscultopercussion test/auscultoscraping test** to find out the greater curvature of the stomach.
   Procedure: Keep the "bell" of stethoscope in the centre of epigastrium (ask the patient to hold the bell of steth) and percuss radially. Percussion over the stomach gives a dull note because of presence of fluid. When the note changes, it indicates the greater curvature of stomach. Mark it on the abdomen (instead of percussion, scraping can be done with finger nail). A line joining 3 or 4 such marks outlines the greater curvature of stomach.

Saline load test: 700 ml of normal saline is infused into stomach over a period of 3.5 minutes through a nasogastric tube and is clamped. If a volume more than 350 ml can be aspirated, it indicates obstruction.

Electrolyte changes in gastric outlet obstruction

- Hypochloraemic alkalosis
- Hyponatraemia
- Hypokalaemia
- Paradoxical aciduria (Fig. 23.55)

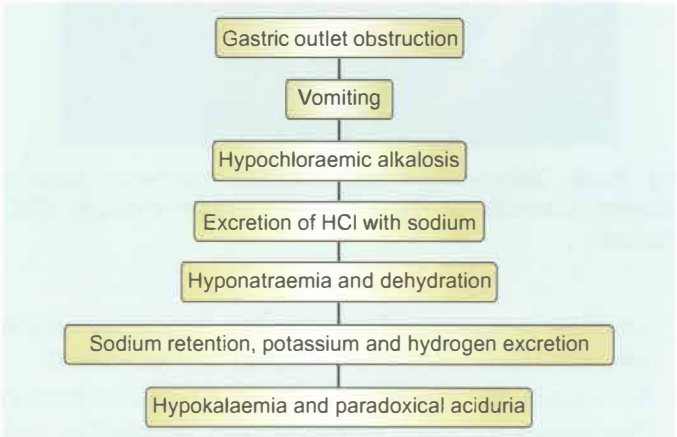


Fig. 23.55: Pathogenesis of paradoxical aciduria

Investigations

1. **Barium meal X-ray**
   - **Hugely dilated stomach (large and low stomach).**
   - Barium does not enter the duodenum. Barium mixed with food residue can give rise to ***mosaic appearance***. Delay in evacuation (on repeat X-ray) of barium into the duodenum is seen (Fig. 23.56).
2. **Gastroscopy**
   - The scope will not enter the duodenum. Stomach is full of foul-smelling food residue.
   - Gastroscopy is also done to rule out carcinoma of the stomach.
3. **Electrolyte study (*vide infra*)**

Treatment (ABCDEF)

A: Aspiration with Ryle's tube—good stomach wash, twice a day is given to keep the stomach empty. Saline is used as it

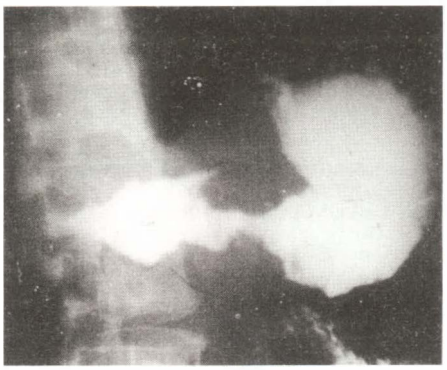


Fig. 23.56: Barium meal showing intrinsic, irregular, persistent filling defect involving pyloric antrum

decreases oedema of the stomach wall. Stomach wash should be given for at least 3–5 days before surgery.

B: Blood is arranged for surgery. Blood may be required preoperatively to treat anaemia.

C: Charts: Adequate urine output is maintained by infusion of intravenous fluids.

D: Drug: Antibiotics for surgery.

E: Exploratory laparotomy: **Vagotomy followed by GJ is done.** Pyloroplasty should not be done because the duodenum is scarred, cicatrised, fibrosed and narrowed.

F: Fluids to correct electrolyte abnormalities. Pyloric stenosis patients can develop **"Hyponatraemic, Hypochloraemic, Hypokalaemic alkalosis"**, Ringer lactate is an ideal supplement.

- Postoperatively these patients recover very fast. Dehydration improves and nutritionally they show dramatic improvement. Even the gastric tone may return after a few years.

Differential diagnosis of gastric outlet obstruction

Carcinoma pyloric antrum (Table 23.8)

II. Tea-pot deformity—handbag stomach (Fig. 23.57)

- A long-standing lesser curve gastric ulcer causes shortening of the lesser curvature due to fibrosis. Such stomach resembles a tea-pot. As a result of this, the pylorus becomes nondependent. Hence, stasis occurs.
- **Treatment:** Partial gastrectomy followed by Billroth I anastomosis.

III. Hourglass contracture (Fig. 23.58)

- When a saddle-shaped ulcer in the lesser curvature gets cicatrised, it involves both surfaces of the stomach resulting in conversion of stomach into two compartments.
- Features of stasis such as fullness, distension and persistent vomiting are present.
- Females are affected more often.
- Weight loss is present. Appetite is decreased.
- It is treated by **Billroth I partial gastrectomy** with removal of 2nd pouch.

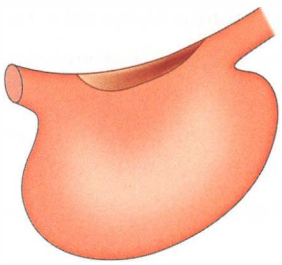


Fig. 23.57: Tea-pot deformity

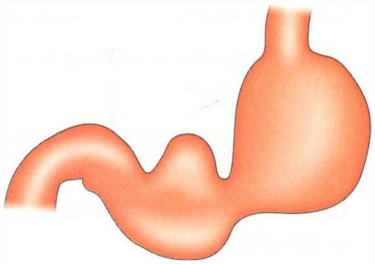


Fig. 23.58: Hourglass contracture of the stomach

IV. Penetration into pancreas

Posterior gastric ulcer can penetrate into pancreas, resulting in severe referred pain to the back resembling pancreatic pathology. However, this type of pain is relieved on lying down.

CARCINOMA STOMACH

Introduction (Clinical notes and Key Box 23.19)

- Carcinoma stomach is more common (2 times) in men compared to women.
- Rare below 40. Average age is 63 years.

Table 23.8 Differential diagnosis of pyloric stenosis

	Cicatrised chronic duodenal ulcer	Carcinoma pyloric antrum
1. Age	20–40 years	> 40–50 years
2. Duration of the symptoms	Long duration of abdominal pain and recent history of vomiting	Pain is usually absent, but vomiting is a feature. Symptoms are of short duration
3. Appetite	Decreased because of vomiting	Severe loss of appetite
4. Weight loss	Present	Significant
5. Anaemia	Not a feature	Present
6. Mass	Not palpable	Hard, irregular mass is palpable

- The palpable mass is the deciding clinical factor (sign). Other differences cannot truly differentiate between the two conditions. Rarely, carcinoma of the stomach is also seen in young patients at the age of 20. In congenital hypertrophic pyloric stenosis, the mass is palpable (page 510).

KEY BOX 23.19

CARCINOMA PROXIMAL STOMACH (CARDIA)

- Incidence is increasing. Obesity and high socioeconomic group
- More aggressive
- Thin muscularis mucosa. Hence, submucosal invasion is seen early
- Diagnosis may also get delayed as endoscopy needs technical expertise
- Signet ring carcinoma is common here
- Surgical resection involves oesophageal anastomosis which is technically demanding. Leak rates are high
- Hence, prognosis is poor

- Incidence of proximal gastric carcinoma is increased—may be due to obesity and in rich socioeconomic status patients.
- Carcinoma distal stomach is more commonly associated with *H. pylori*.
- Proximal carcinomas are more advanced at the time of presentation than distal carcinomas (Fig. 23.59).
- Overall 5-year survival after the diagnosis of gastric cancer is 10 to 20%.
- Those who undergo potentially curative resection (R–0) have a 5-year survival rate of 25–50%.

CLINICAL NOTES

A 32-year-old male was admitted with loss of appetite of 3 months duration. Endoscopy revealed a growth in the body of the stomach. At exploration, large para-aortic nodes were present. Subtotal gastrectomy was done. He died after 6 months due to extensive metastasis. No wonder, carcinoma of the stomach is called ***Captain of men of death.***

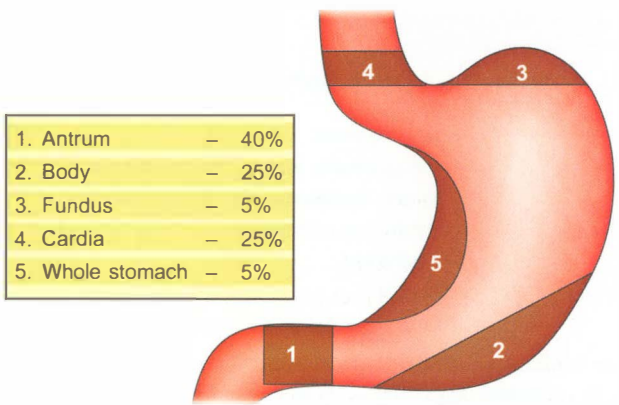


Fig. 23.59: Site and incidence of gastric cancer

Risk factors for carcinoma stomach

(Fig. 23.60 and Key Box 23.20)

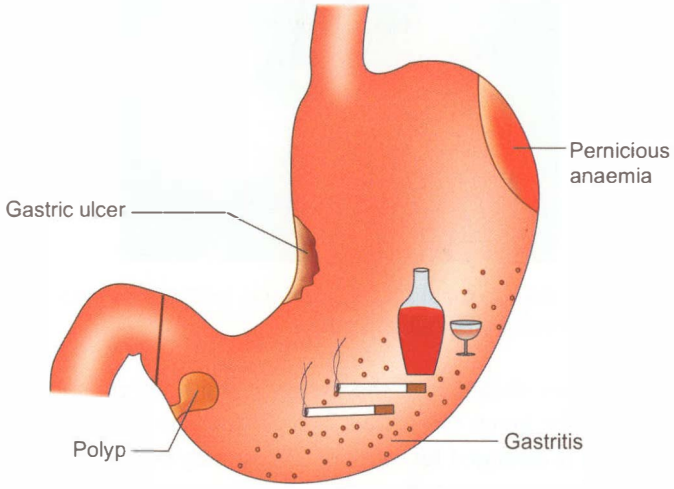


Fig. 23.60: Risk factors for carcinoma stomach

1. Environmental and dietary factors

- The incidence is increased in persons who consume red meat, cabbage, spices, spirits, salt-fish (Key Box 23.20).
- Smoked salmon fish was responsible for increased incidence of carcinoma stomach in Japanese population. Probably, it is related to release of polycyclic hydrocarbons and aromatic amino acids. Smoking, spicy food and alcohol consumed over a period of many years produce chronic gastritis which may change into carcinoma of stomach.

KEY BOX 23.20

FOOD PRODUCTS WHICH MAY BE CARCINOGENIC

- Smoked food
- Spirits
- Smoking
- Salted food
- Soil
- Diet low in carbohydrate
- Animal protein
- Rich fatty food
- High in complex carbohydrates
- *H. pylori* in contaminated water

PEARLS OF WISDOM

WHO recommends increased consumption of fruits and vegetables. Vitamin C, is an antioxidant which is a protective agent. Ascorbic acid prevents conversion of nitrates to nitrites.

2. Precancerous conditions

a. **Atrophic gastritis:** This may be due to smoking, spicy food, continuous ingestion of drugs, reflux of bile into the stomach, etc.

b. Pernicious anaemia: Patients have increased risk (four to six times) of development of carcinoma when compared to general population.

- It causes atrophic gastritis and precipitates carcinoma of fundus of the stomach. Lesions are polypoidal and multicentric

c. Patients with **hypogammaglobulinaemia** (50-fold increase) are at high-risk.

d. *H. pylori* infection results in atrophic gastritis, followed by the intestinal type of gastric mucosa, metaplasia and then dysplasia. Eventually it leads to **intestinal type of gastric cancer** (Figs 23.61 and 23.62). *H. pylori* can also cause proliferation of gastric cancer cells and decrease secretion of vitamin C. Cytotoxin associated gene A (Cag A) is associated with increased risk.

- Also both **type A** and **type B** gastritis can predispose to carcinoma stomach. Type A—proximal stomach, type B—distal stomach.

e. Adenomatous polyps which occur in the antrum have highest risk of malignant transformation (larger polyps, i.e. more than 2 cm—10 to 20% malignant transformation).

- Polyp more than 2 cm, pedunculated polyp can be removed by endoscopically. Higher chances of malignancy is seen in sessile polyps.

f. Menetrier's disease is a protein-losing enteropathy, along with giant hypertrophy of gastric mucosal folds. It is a precancerous condition.

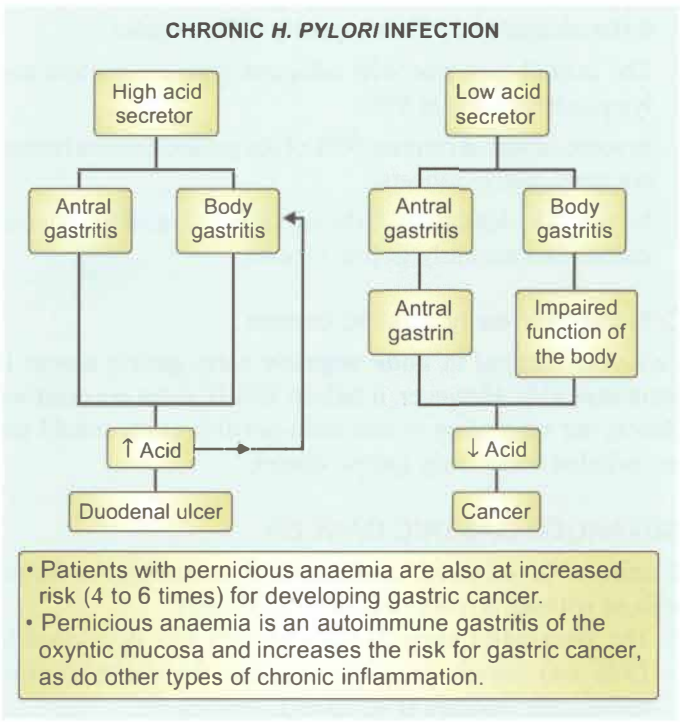


Fig. 23.61: *H. pylori* and pathogenesis of duodenal ulcer and carcinoma stomach

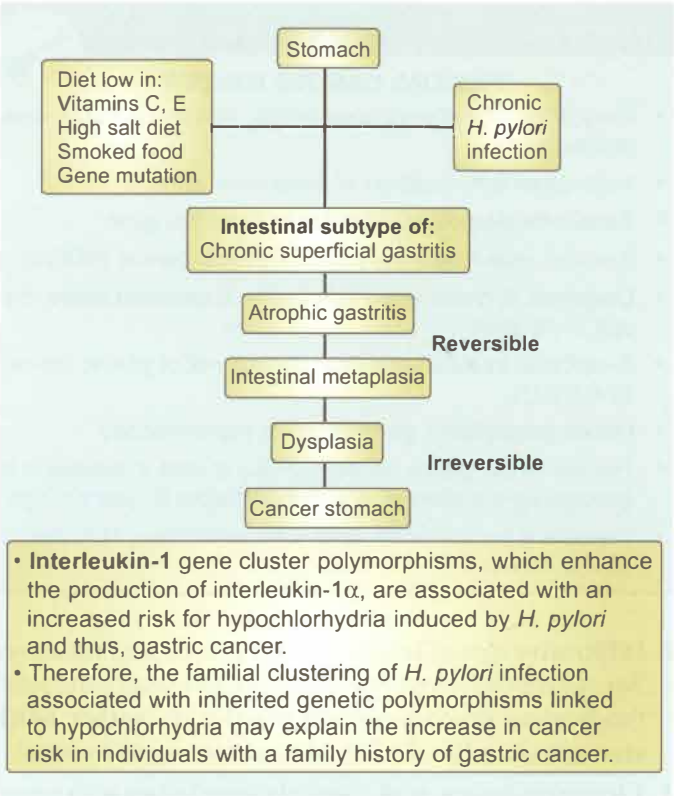


Fig. 23.62: Aetiopathogenesis of carcinoma stomach

g. Gastric ulcer (benign): Incidence of malignancy is 2% (0.5 to 5%). Carcinoma arising in a gastric ulcer is called "*Ulcer Cancer of the Stomach*".

h. Previous GJ or gastric resection predisposes to development of carcinoma of the stomach after a period of 15–20 years. Such a carcinoma is described as stump carcinoma. Pathogenesis is related to development of atrophic gastritis, achlorhydria and duodenogastric bile reflux.

3. Genetic and familial factors (Key Box 23.21)

- Carcinoma stomach can run in families. However, only 10% of patients give family history of carcinoma stomach.
- Carcinoma stomach is more common in patients with blood group A. These patients have different mucopolysaccharide secretion in the stomach and greater susceptibility to ingested carcinogens. They develop diffuse type of carcinoma.
- Several genetic alterations have been identified such as activation of oncogenes, inactivation of tumour suppressor genes p53 and p16, reduction or loss in the cell adhesion molecule E-cadherin (met proto-oncogene).

Gross types

1. **Cauliflower** like growth with friable tissue. This variety can give rise to melaena or bleeding causing anaemia.

KEY BOX 23.21

FAMILIAL GASTRIC CANCER

- Only 10% of patients give family history of carcinoma stomach
- Associated with mutation of E-cadherin gene
- Bonaparte (Napoleon) appears to have this gene
- It is also called hereditary diffuse gastric cancer (HDGC)
- Diagnosis is made when carcinoma is detected below the age of 40 years
- E-cadherin mutation results in lifetime risk of gastric cancer by 60–90%.
- Hence prophylactic gastrectomy is recommended
- Familial diffuse gastric cancer—(FGC) at least 3 relatives in 2 generations are affected, one of them before 50 years of age.
- Hereditary nonpolyposis colon cancer carries—10% risk of gastric cancer

2. **Infiltrative** type of lesion (diffuse) with dense submucosal fibrosis which converts the stomach into a small contracted, functionless stomach—**linitis plastica** or **leather bottle stomach** (Key Box 23.22). Mucosa may appear normal.
3. **Ulcerative** variety, with classical everted edges with central slough.
4. **Ulcer cancer** refers to carcinoma arising in a pre-existing gastric ulcer. In this variety, complete destruction of the muscle coat is present.
5. **Colloid carcinoma:** In this condition, malignant cells are separated by colloid material. This is the type which is common in women and gives rise to Krukenberg's tumour—bilateral, bulky ovarian metastasis common in premenopausal women (signet ring carcinoma produces this).

Pathology

- Ninety-five percent of all malignant gastric neoplasms are adenocarcinomas.
- The Lauren system separates gastric adenocarcinoma into intestinal or diffuse types based on histology (Key Box 23.23).
- Other histologic types include squamous cell carcinoma, adenoacanthoma, carcinoid tumours, GI stromal tumours, and lymphoma.

EARLY GASTRIC CANCER (Key Box 23.23)

- Early gastric cancer is defined as adenocarcinoma limited to the mucosa and submucosa of the stomach, regardless of lymph node status.
- The entity is common in Japan, where gastric cancer is the number one cause of cancer death, and where aggressive surveillance programs have been established.
- Approximately 10% of patients with early gastric cancer will have lymph node metastasis.

KEY BOX 23.22

EARLY GASTRIC CANCER (JAPANESE CLASSIFICATION)

Type I Exophytic lesion extending into the gastric lumen
Type II Superficial variant
- **II A** Elevated lesions with a height no more than the thickness of the adjacent mucosa
- **II B** Flat lesions
- **II C** Depressed lesions with an eroded but not deeply ulcerated appearance

Type III Excavated lesions that may extend into the muscularis propria without invasion of this layer by actual cancer cells

KEY BOX 23.23

LAUREN CLASSIFICATION SYSTEM

INTESTINAL	DIFFUSE
• Environmental	Familial, blood type A
• Gastric atrophy, intestinal metaplasia	Does not arise in gastritis
• Men > women	Women > men
• Higher incidence with age	Younger age group
• Gland formation present	Poorly differentiated, signet ring cells, no gland formation
• Haematogenous spread	Transmural/lymphatic spread
• Microsatellite instability	
APC gene mutations	Decreased E-cadherin
p53, p16 inactivation	*p53, p16* inactivation

- Approximately 70% of early gastric cancers are well differentiated and 30% are poorly differentiated.
- The overall cure rate with adequate gastric resection and lymphadenectomy is 95%.
- In some Japanese centres, 50% of the gastric cancers treated are early gastric cancers.
- In the US, less than 20% of resected gastric adenocarcinomas are early gastric cancers.

Criticism for early gastric cancer

Five-year survival in **node negative** early gastric cancer is more than 95%. However, it falls to 70% if nodes are positive. Hence, the suggestion is that node-positive cases should not be included under early gastric cancer.

ADVANCED GASTRIC CANCER

It refers to involvement of muscularis mucosa and/or serosa with or without involvement of lymph nodes.

- The **Borrmann classification system** was developed in 1926 and remains useful today for the description of endoscopic findings (Fig. 23.63).
- The Borrmann system divides gastric carcinoma into five types depending on the lesion's **macroscopic** appearance.

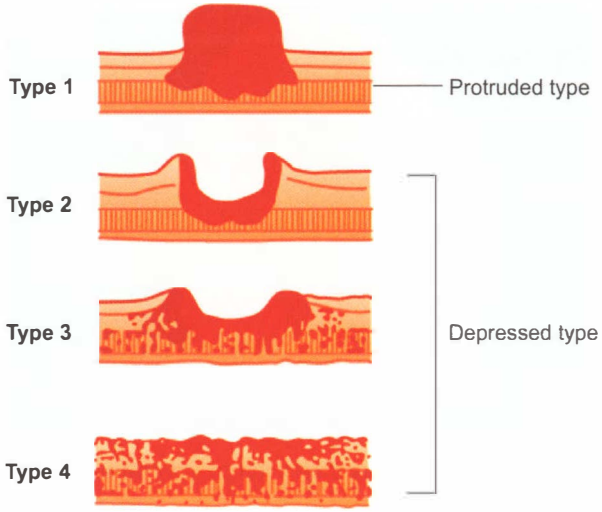


Fig. 23.63: Borrmann's classification (*refer to* advanced gastric cancer macroscopic type)

- **Type 1** represents polypoid or fungating lesions
- **Type 2** ulcerating lesions surrounded by elevated borders
- **Type 3** ulcerating lesions with infiltration into the gastric wall
- **Type 4** diffusely infiltrating lesions
- **Type 5** lesions that do not fit into any of the other categories.

- ***Linitis plastica*** is the term to describe type 4 carcinoma when it involves the entire stomach (Key Box 23.24 and Figs 23.64 to 23.66).

KEY BOX 23.24

THE DIFFUSE FORM OF GASTRIC ADENOCARCINOMA—LINITIS PLASTICA

- **Poorly differentiated**
- Lacks gland formation
- Composed of **signet ring** cells
- Consists of tiny clusters of small uniform cells
- Tends to spread submucosally
- Has less inflammatory infiltration
- **Metastases early**
- Route of spread is generally by transmural extension and through lymphatic invasion
- Does not generally arise in the setting of prior gastritis
- **More common in women**
- Affects a slightly younger age group
- Association with blood type A and familial occurrences, suggesting a **genetic aetiology**
- Intraperitoneal metastases are frequent
- The prognosis is less favourable for patients with diffuse-subtype histology.

Fig. 23.64: Localised linitis plastica

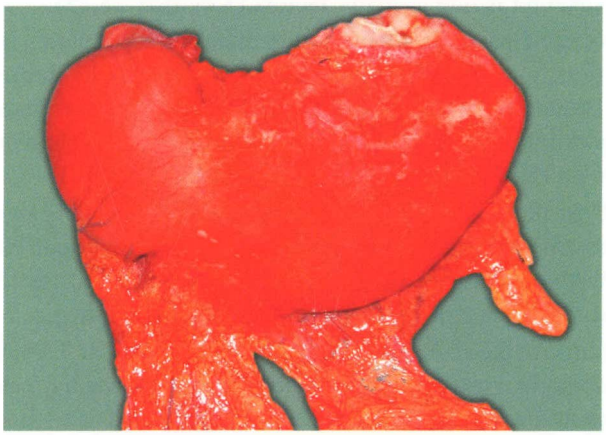


Fig. 23.65: Diffuse variety of carcinoma stomach

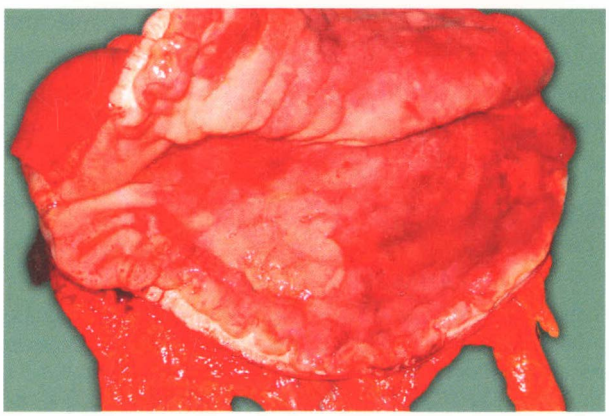


Fig. 23.66: Linitis plastica—opened specimen

- The original histologic classification system was developed by Borders in 1942.
- Borders classified gastric carcinomas according to the degree of cellular differentiation, independent of morphology, and ranged from 1 (well-differentiated) to 4 (anaplastic).

WHO CLASSIFICATION—five main categories

1. Adenocarcinoma 95%
   - Papillary, tubular
   - Mucinous
   - Signet ring
2. Adenosquamous cell carcinoma
3. Squamous cell carcinoma
4. Undifferentiated carcinoma
5. Unclassified carcinoma

Clinical features of carcinoma stomach (SOLID)[1]

- Very often patients would have vague symptoms—early satiety, flatulence, discomfort, pain in the upper abdomen.
- Early satiety is due to decreased distensibility of the stomach (Fig. 23.67).
- Anaemia is due to many factors (Key Box 23.25).

S **S**ilent: Growth is silent but manifests as secondaries in the liver, ascites, Virchow's node, rectovesical deposits, (Blumer's shelf), umbilical nodule (Sister Mary Joseph's nodule), left axillary nodes (Irish nodes), palpable ovarian mass (Krukenberg tumour) (Figs 23.67 to 23.72).

O **O**bstruction at pylorus (pyloric antrum) producing pyloric obstruction with features of vomiting with/without blood. Visible gastric peristalsis can also be present. Obstruction at cardio-oesophageal junction produces dysphagia.

L **L**ump in the abdomen which is hard and irregular. Clinically, stomach mass is differentiated from liver mass by features mentioned below.

I **I**nsidious in onset: Anaemia, anorexia and asthenia of short duration.

D **D**yspepsia in a man over the age of 40: Carcinoma stomach should be ruled out. Early gastric cancer presents as dyspepsia.

Nonmetastatic conditions such as thrombophlebitis (Trousseau's sign) and deep venous thrombosis can occur due to change in thrombotic and haemostatic mechanisms.

KEY BOX 23.25

ANAEMIA IN CARCINOMA

- This is one of the common presentations. Often patients get investigated for anaemia by the physician only to discover carcinoma stomach.
- Achlorhydria results in poor conversion of ferrous to ferric which causes anaemia.
- 15% of patients may also develop haematemesis as in ulcerative lesions or proliferative lesions. GI blood loss also accounts for anaemia.
- Early satiety, loss of appetite and poor intake also contribute to anaemia (minor role).
- 40% of carcinoma stomach patients have anaemia.

[1]If you remember **SOLID**, you will get solid marks.

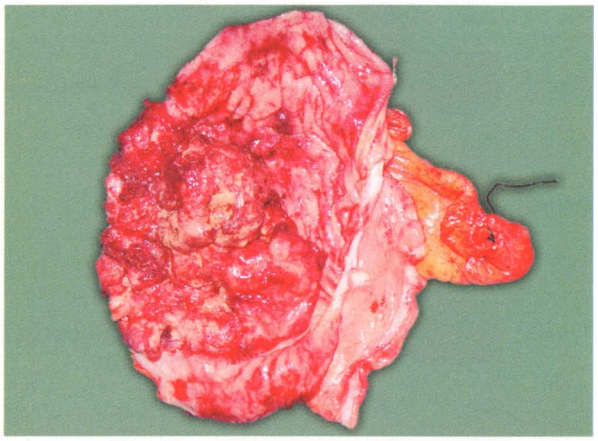


Fig. 23.67: Carcinoma stomach large ulcerative lesion with involvement of serosa

Fig. 23.68: Umbilical nodule—popularly called Sister Mary Joseph's nodule

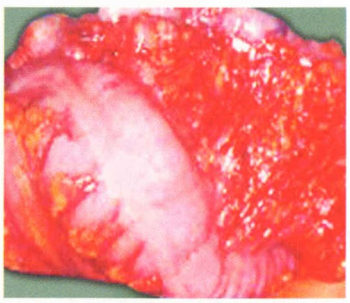


Fig. 23.69: Carcinoma stomach with nodules in the greater omentum

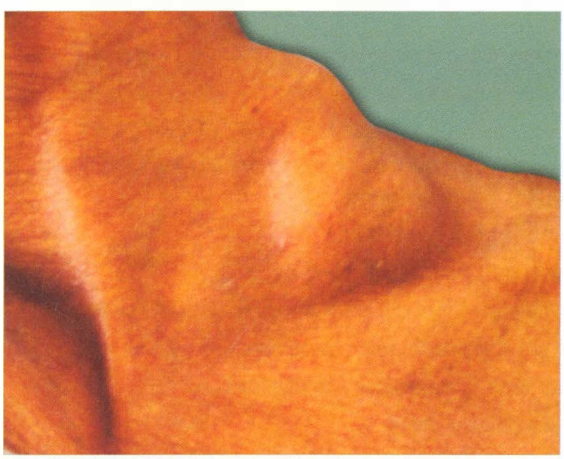


Fig. 23.70: Large supraclavicular nodes

Features of stomach mass

1. Stomach moves with respiration.
2. Upper border of the stomach mass can be made out.
3. Anatomical location of the mass: Right hypochondrium in a pyloric mass, epigastrium and left hypochondrium in a mass arising from body of the stomach.
4. Knee elbow position: The mass falls forwards, unless fixed.
5. The mass may have intrinsic mobility.

Fig. 23.71: Krukenberg tumours

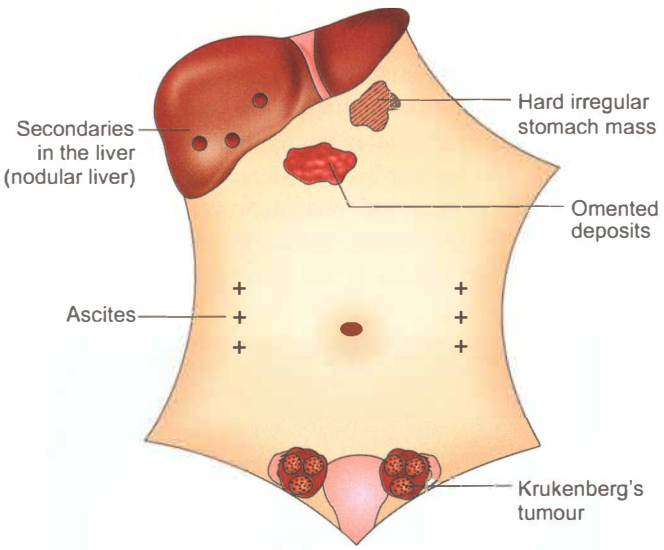


Fig. 23.72: Diagrammatic representation of secondaries in carcinoma stomach

Spread

1. **Penetration of gastric serosa:** This is the most important prognostic indicator. When serosa is NOT penetrated, 50% survive for 5 years after resection. When serosa is penetrated, this figure drops to 20%.
   - Once serosa is involved, adjacent organs such as liver, pancreas, spleen, omentum, transverse colon get involved.
2. **Lymphatic spread:** 420 lymph nodes have been identified
   - Lymph node involvement is a poor prognostic indicator.
   - Involvement of 4 or more nodes is less favourable.

*Staging refer TNM staging

Lymphatic zones

Lymphatic drainage from the stomach has been classified into four zones.

- **Zone 1:** In the gastrocolic omentum along the right gastroepiploic vessels. This drains the pyloric antrum and lower half of greater curvature.
- **Zone 2:** It lies in the gastrocolic omentum and gastrosplenic ligament along the left gastroepiploic vessels. This drains upper half of the greater curvature.
- **Zone 3:** It lies in the lesser omentum draining proximal two-thirds of the stomach. From here, lymph drains into perioesophageal lymph nodes.
- **Zone 4:** It is from distal portion of the lesser curve and pylorus along hepatic artery and right gastric artery into para-aortic nodes.

3. **Blood spread:** Most common sites are liver and lungs. It produces extensive secondaries. They are signs of inoperability.
4. **Transcoelomic spread** results in ascites, Krukenberg tumour—bilateral bulky ovarian deposits and rectovesical deposits **(Blumer's shelf)**.

TNM STAGING

AJCC TNM staging

Primary tumour (T)

TX	Primary tumour cannot be assesed
T0	No evidence of primary tumour
Tis	Carcinoma *in situ*. Intraepithelial tumour without invasion of the lamina propria
T1	Tumour invades lamina propria, muscularis mucosa or submucosa
T1a	Tumour invades lamina propria or muscularis mucosa
T1b	Tumour invades submucosa
T2	Tumour invades muscularis propria
T3	Tumour penetrates subserosal connective tissue without invasion of visceral peritoneum or adjacent structures
T4	Tumour invades serosa (visceral peritoneum) or adjacent structures
T4a	Tumour invades serosa (visceral peritoneum)
T4b	Tumour invades adjacent structures

Regional lymph nodes (N)

NX	Regional lymph nodes (S) cannot be assessed
N0	No regional lymph node metastasis($)
N1	Metastasis in 1–2 regional lymph nodes
N2	Metastasis in 3–6 regional lymph nodes
N3	Metastasis in 7 or more regional lymph nodes
N3a	Metastasis in 7 to 15 regional lymph nodes
N3b	Metastasis in 16 or more regional lymph nodes

Distant metastasis (M)

M0	No distant metastasis
M1	Distant metastasis

Histologic grade (G)

GX	Grade cannot be assessed
G1	Well-differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated

Investigations

1. **Complete blood picture:** 20% of early gastric cancer patients have iron deficiency (microcytic, hypochromic) anaemia. Preoperative blood transfusion may be necessary.
2. Routine examination, fasting and postprandial sugars, ECG, renal function for fitness before surgery.
3. **Flexible oesophagogastroduodenoscopy** (Figs 23.73 to 23.76)

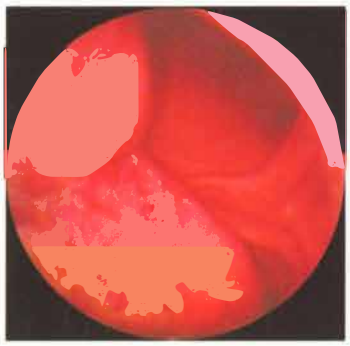


Fig. 23.73: Endoscopy showing superficial ulceration

Fig. 23.74: Endoscopy showing ulceroproliferative growth

Fig. 23.75: Friable granular tissue—antral carcinoma

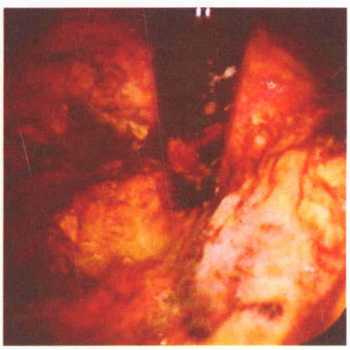


Fig. 23.76: Fundal carcinoma detected by J manoeuvre in endoscopy

- To know the extent of the lesion
- To confirm the diagnosis
- To take multiple biopsy—6 pieces
- Also to aid brush cytology

4. **Ultrasound and CT scan**
   - To rule out secondaries in the liver.
   - To look for enlarged coeliac nodes.
   - Can detect ascites—guided fluid tap and cell cytology.
   - To detect Krukenberg tumour (pelvic CT).
   - Useful in detecting metastatic disease.
5. **Endoscopic ultrasonography** can differentiate early gastric cancer from advanced tumours in 80% of patients. Overall staging accuracy is about 75%.
6. **Laparoscopy:** CT cannot detect liver or peritoneal metastasis (small < 5 mm) and small lymph nodes. Laparoscopy is an ideal investigation. Almost 20 to 30% of so-called operable cases become inoperable. Laparoscopic peritoneal lavage for cytology is best test.
7. **Barium meal** may show intrinsic, persistent, irregular, filling defect. Double contrast air-barium study is used for mass screening in Japan to detect early cases (Figs 23.77 to 23.79)
   - Barium meal study is **useful in cases of linitis plastica** wherein mucosa may appear to be normal in early cases
   - Today use of barium has become almost nil with the availability of endoscopy.
8. **CEA:** Carcinoembryonic antigen is elevated in about 60–70% patients. It indicates extensive spread of the disease.

Histopathology

- It is an adenocarcinoma of the stomach. There are basically two types of gastric carcinomas as per Lauren's classification.
- **D**iffuse is more common in young, females and carries poor prognosis. The leather-bottle stomach or linitis plastica is poorly differentiated with anaplastic cells.
- **I**ntestinal is more common in elderly males. It shows areas of intestinal metaplasia. It has better prognosis.

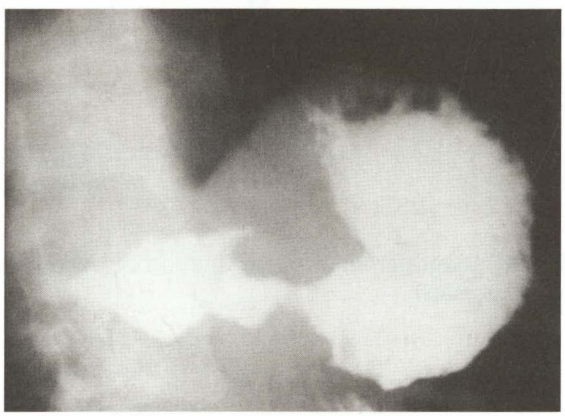


Fig. 23.77: Barium meal study showing filling defect

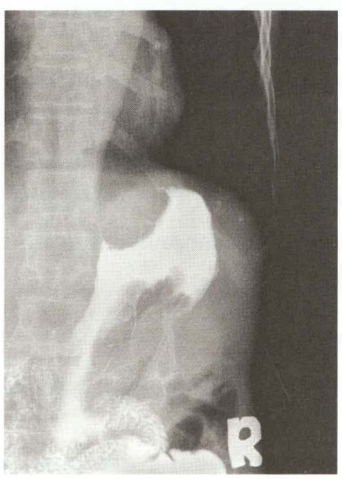


Fig. 23.78: Carcinoma stomach—Barium meal showing a large irregular filing defect along the greater curvature

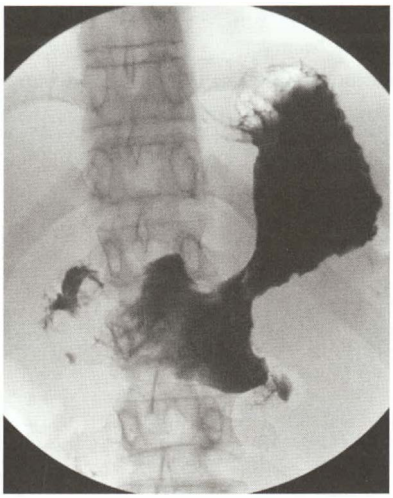


Fig. 23.79: Hour glass contracture of the stomach caused by carcinoma

Treatment of carcinoma stomach

- **Surgery is the main modality of the treatment.** Adjuvant chemotherapy has been found to be beneficial in a few patients only.
- Resectable means the growth can be removed.
- Inoperable means there are no chances of cure but growth may be resectable. Operable means cure is possible.

Signs of inoperability

- Growth fixed to pancreas or posterior abdominal wall
- Secondaries in the liver, hard nodular liver
- Rectovesical deposits, due to peritoneal seedings which are felt during per rectal examination
- Enlarged, fixed coeliac nodes, para-aortic nodes and left supraclavicular nodes
- Krukenberg tumour, malignant ascites
- Sister Mary Joseph's nodule

SISTER MARY JOSEPH NODULE

- Sister Mary Joseph—head nurse to William Mayo
- Was first to notice that a 'nodule' in the umbilicus was often associated with advanced malignancy
- Presents as firm, red, nontender nodule
- Results from spread of tumour within the falciform ligament
- 90% of tumours are adenocarcinomas
- Commonest primaries are stomach and ovary
- Primary tumour is almost invariably inoperable

Aims of surgery

1. Curative resection should be done whenever possible
2. Bypass procedure (GJ) to relieve vomiting in advanced cases of pyloric obstruction.
3. Palliative gastrectomy can be done to remove a fungating, ulcerative, bleeding mass. It gives better palliation.

Curative resections

A resection is considered to be curative if

- There is no evidence of microscopic or gross residual tumour.
- Serosa is not involved (this means that curative resection is not possible for T3/T4 tumours).
- There is no evidence of metastatic disease.

Terminology

- Japanese Research Society for gastric cancer advocates very aggressive resection including lymphadenectomy (Fig. 23.80). Hence, more details of lymph node station are given in Table 23.9. However, the rest of the cancer research groups were not able to produce the same results as the Japanese.
- **D1** resection refers to the removal of primary group of nodes such as nodes along the lesser and greater curvature, and juxtapyloric nodes. **This will become curative resection when lymph nodes are clinically not enlarged (N0) but they are removed** (Fig. 23.81).
- **D2** resection refers to the removal of lymph nodes such as left gastric, common hepatic, splenic, retropancreatic nodes, etc. **This will become curative resection when lymph nodes are clinically enlarged (N1)** (Key Box 23.26 and Fig. 23.82).
- **D3** resection refers to the removal of lymph nodes such as para-aortic, porta hepatis nodes, behind the head of the pancreas, etc. (not done).

D resection exceeds the nodal involvement by one level.
N0—D1 resection is curative
N1—D2 resection is curative and so on.

Extent of gastric resection

- The extent of the gastrectomy is site-dependent and focuses on complete removal of the gastric carcinoma with preferably a 6 cm margin from the gross edge of the tumour.

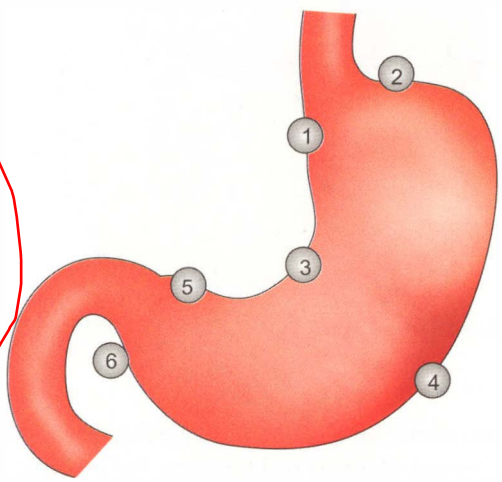


Fig. 23.80: Perigastric nodes: (1) right pericardial; (2) left pericardial; (3) lesser curvature; (4) greater curvature; (5) suprapyloric; (6) infrapyloric

Table 23.9 Grouping of regional lymph nodes

Lymph node station (number)	Description	Location of primary tumour in the stomach		
		Upper third	Middle third	Lower third
1	Right paracardial	1	1	2
2	Left paracardial	1	3	M
3	Lesser curvature	1	1	1
4sa	Short gastric	1	3	M
4sb	Left gastroepiploic	1	1	3
4d	Right gastroepiploic	2	1	1
5	Suprapyloric	3	1	1
6	Infrapyloric	3	1	1
7	Left gastric artery	2	2	2
8a	Anterior common hepatic	2	2	2
8p	Posterior common hepatic	3	3	3
9	**Coeliac artery**	2	2	2
10	**Splenic hilum**	2	3	M
11p	Proximal splenic	2	2	2
11d	Distal splenic	2	3	M
12	Hepatoduodenal	3	2	2
13	**Retropancreatic**	M	3	3
14v	Superior mesenteric vein	M	3	2
14a	Superior mesenteric artery	M	M	M
15	Middle colic	M	M	M
16a1	Aortic hiatus	3	M	M

Numbers 1–3: Group number M = Lymph nodes regarded as distant metastasis

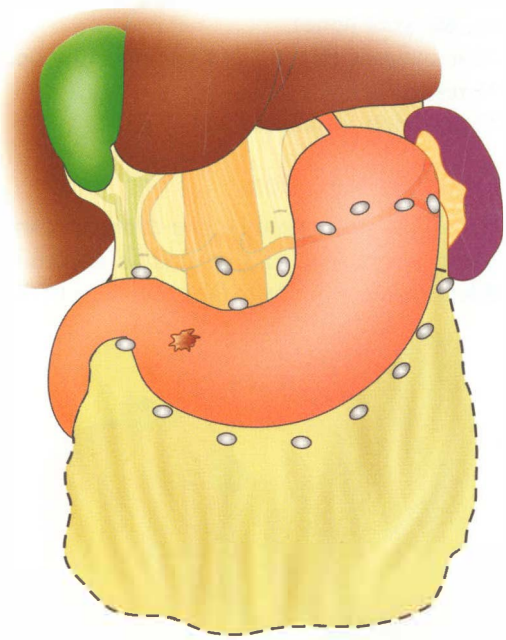


Fig. 23.81: Extent of D1 resection of tumour in distal one-third of stomach

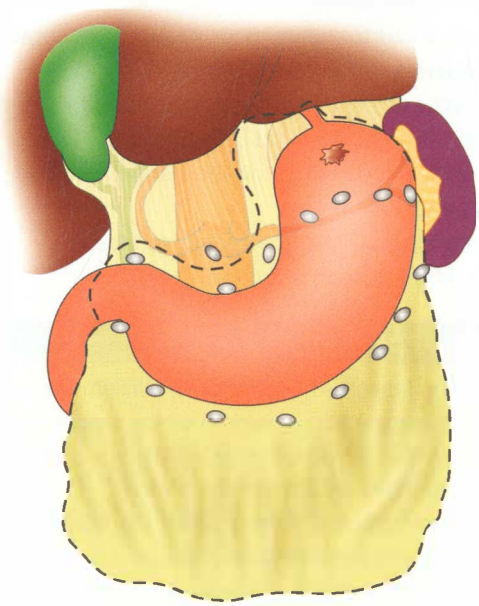


Fig. 23.82: D2 resection of tumours of the proximal third of stomach

- Clearly, anatomic limitations influence this margin because in antral lesions close to or involving the pylorus, only a limited portion of the duodenum can be removed.
- In patients with a distal lesion, essentially a distal subtotal gastrectomy is performed regardless of T stage.

- **For proximal gastric cancers (fundic),** total gastrectomy is required.
- **For midbody or more extensive lesions,** total gastrectomy is required.

OPERATIVE PHOTOGRAPHS OF CARCINOMA STOMACH (Figs 23.83 to 23.88)

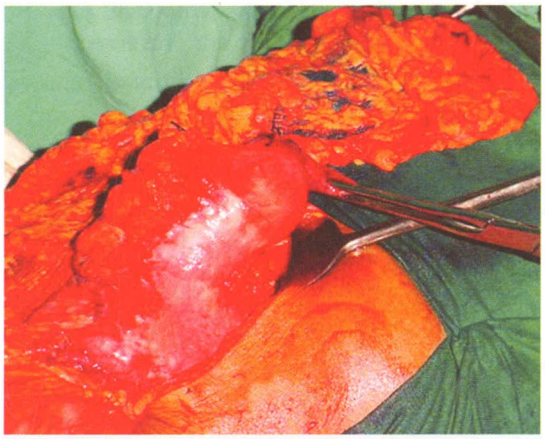


Fig. 23.83: Carcinoma stomach with involvement of serosa, endosonogram can detect this

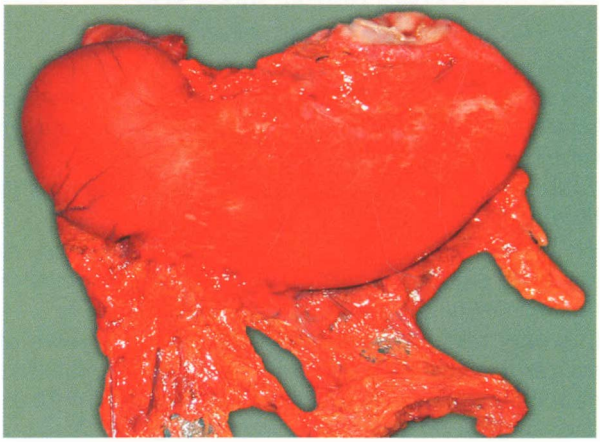


Fig. 23.84: Total gastrectomy—diffuse carcinoma of the stomach, known for transmural spread

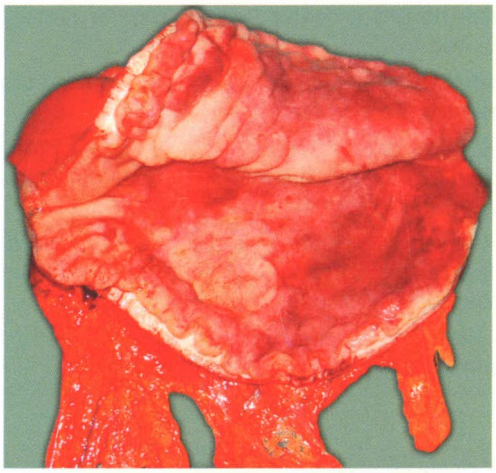


Fig. 23.85: Opened specimen—loss of rugae, nodular cancerous tissue

Fig. 23.86: Subtotal gastrectomy for antral carcinoma stomach. They present with gastric outlet obstruction

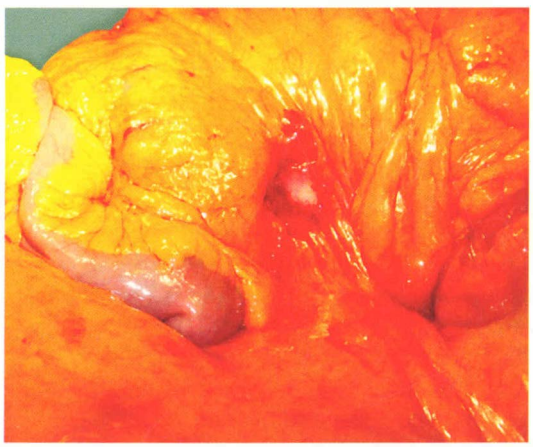


Fig. 23.87: Peritoneal metastasis from carcinoma stomach. Laparoscopy is the most ideal investigation to detect peritoneal metastasis

Fig. 23.88 Intestinal type—gland formation present, similar to adenocarcinoma colon—good prognosis

KEY BOX 23.26

PATHOLOGICAL BASIS FOR D2 DISSECTION

- Gastric cancer has a great propensity to spread through the rich plexus of lymphatics of the stomach to local or regional lymph nodes. Hence, the need to remove stomach and lymph nodes.
- **Gastric cancer, unlike breast cancer, remains for a long time as a locoregional disease**.
- It is striking too, that when gastric cancer recurs it often does so locoregionally rather than more wide-spread dissemination.
- It is very rare for gastric cancer to recur 5 years after surgery, whereas for breast cancer, disseminated micrometastases continue to take their toll some 10–20 years after surgery.

- **For more distal lesions,** a subtotal gastrectomy is the preferred approach.
- **Extended organ resection** is reserved for patients with apparently node-negative T4 lesions, in which complete resection requires resection of the invaded portions of the diaphragm, pancreas, spleen, adrenal gland or colon.
- These patients usually are pretreated with chemotherapy.

Extent of lymphadenectomy

- Generally these are grouped into level N1 (i.e. stations 3 to 6),
- Level N2 (i.e. stations 1, 2, 7, 8 and 11) and
- Level N3 (i.e. stations 9, 10 and 12) nodes.
- The nodal stations defined as level N1, N2, and N3 vary depending on the location of the tumour.

PEARLS OF WISDOM

- ***In general, N1 nodes are within 3 cm of the tumour.***
- ***N2 nodes are along the hepatic and splenic arteries.***
- ***N3 nodes are the most distant.***

Extent of gastric and lymph node resection, upper one-third lesions

- A tumour is considered resectable if confined to stomach or N1 or N2 nodes are involved
- Nodes less than 3 cm from tumour = N1 nodes
- Nodes greater than 3 cm from tumour = N2 nodes
- If tumour and N1 nodes (Group 1) resected = D1 gastrectomy
- If tumour and N2 nodes (Group 2) resected = D2 gastrectomy
- D3 resection is a D2 resection plus removal of para-aortic lymph nodes.

A standard D2 resection for gastric cancer involves

1. Removing part or whole of the stomach.
2. The N1 (groups 1–6) and N2 (groups 7–11) lymph nodes.
3. The greater and lesser omentum.
4. The spleen and tail of the pancreas for tumours of the proximal stomach in order to remove groups 10 and 11 lymph nodes. Otherwise, they are preserved.
5. D2 resections necessitate the additional removal of the omental bursa.
6. The hepatoduodenal nodes in antral tumours.
7. The splenic artery nodes

R-resections (Figs 23.89 to 23.91)

- The term R status was first described by Hermanek in 1994 and is used to describe the tumour status after resection (Key Box 23.27).

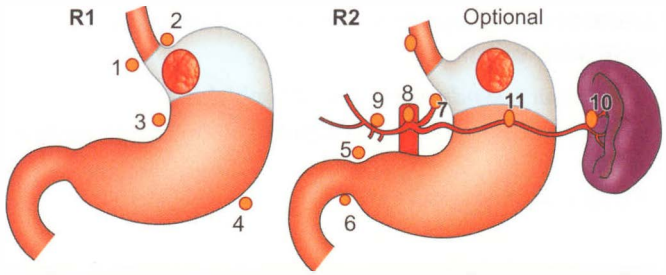
KEY BOX 23.27

R-RESECTIONS

- R0 describes a microscopically margin-negative resection, in which no gross or microscopic tumour remains in the tumour bed.
- R1 indicates removal of all macroscopic disease but microscopic margins are positive for tumour.
- R2 indicates gross residual disease. Since the extent of resection can influence survival, some authors include this R designation to complement the TNM system.
- Long-term survival can be expected only after a R0 resection; therefore, a significant effort should be made to avoid R1 or R2 resections.
- If no disease is identified in the lymph nodes, (N0) nomenclature is used.

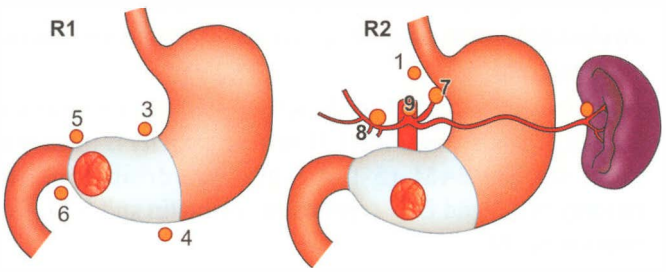
KEY POINTS AND SPECIAL NOTE TO STUDENTS

- Endoscopy has revolutionised the diagnosis of carcinoma stomach. Early diagnosis should be the aim since early carcinoma stomach is curable.
- Ultrasonogram/CT scan are the investigations to detect intra-abdominal metastasis.
- Endosonogram detects various layers of the stomach which are involved by the tumour.
- Diagnostic laparoscopy—it detects peritoneal metastasis—no role for radical surgery.
- Resection is the best line of treatment which can be curative in a few patients.
- Any radical surgery will involve wide excision of the tumour with lymph nodes and omentum.
- Because of added morbidity and a very small survival advantage of D2 resections over D1 resections, many still consider D1 resection as adequate.
- As far as we are concerned, an understanding of these various terminologies and their concepts may enable you to score better marks in theory papers.
- A simple summary of surgical procedures (what everyone should know) has been given here.



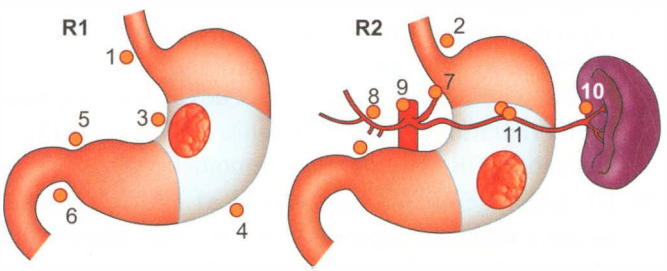
1. Right cardiac	5. Suprapyloric	10. Splenic hilar
2. Left cardiac	6. Infrapyloric	11. Splenic
3. Lesser curvature	7. Left gastric	
4. Greater curvature (and short gastric)	8. Hepatic	
	9. Coeliac	

Fig. 23.89: Proximal gastric cancer



1. Right paracardiac	5. Suprapyloric	8. Hepatic
3. Lesser curvature	6. Infrapyloric	9. Coeliac
4. Greater curvature	7. Left gastric	

Fig. 23.90: Distal gastric cancer



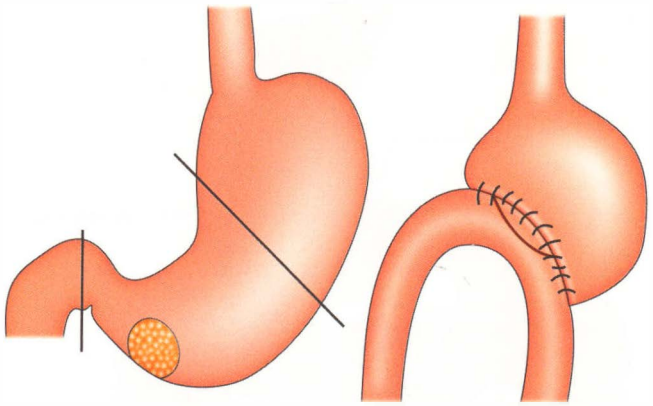
1. Right paracardiac	5. Suprapyloric	9. Coeliac
2. Left paracardiac	6. Infrapyloric	10. Splenic hilar
3. Lesser curvature	7. Left gastric	11. Splenic
4. Greater curvature	8. Hepatic	

Fig. 23.91: Mid-body gastric cancer

Summary of treatment of carcinoma stomach

1. Carcinoma of pyloric antrum and distal body of the stomach

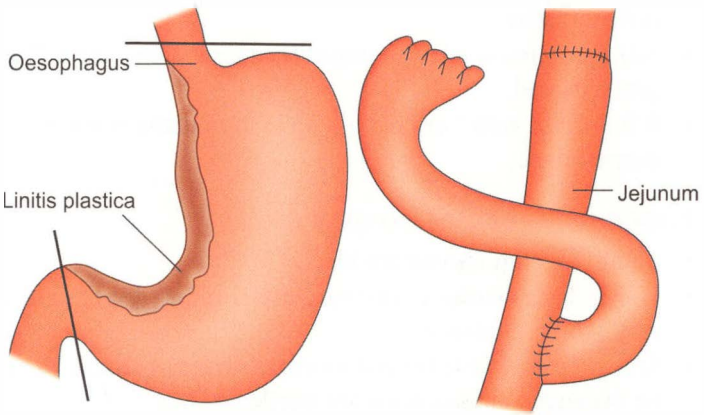
Radical subtotal gastrectomy which includes the removal of 60–70% of the stomach, greater omentum along with enlarged lymph nodes (N1) followed by gastrojejunal anastomosis is the treatment of choice (Figs 23.92 and 23.93).



Figs 23.92 and 23.93: Radical subtotal gastrectomy with Billroth II anastomosis

2. Carcinoma of proximal stomach and diffuse carcinoma

Oesophagogastrectomy (Figs 23.94 and 23.95): Removal of the entire stomach, lower end of oesophagus, with regional lymph nodes, followed by oesophagojejunal anastomosis.



Figs 23.94 and 23.95: Radical total gastrectomy, with Roux-en-Y oesophagojejunostomy

3. Palliative surgery (Fig. 23.96)

1. Carcinoma pyloric antrum (inoperable): Palliative anterior GJ is done to relieve vomiting, by anastomosing a jejunal loop to the stomach in the prepyloric region. If posterior GJ is done, the growth may involve the GJ stoma early resulting in stomal obstruction. With anterior GJ, entero-enterostomy can be added to prevent bilious vomiting.
2. Palliative gastrectomy to get rid of ulcerated, necrotic or bleeding lesion.

Endoscopic mucosal resection

- This is indicated in early gastric cancer confined to mucosa. The cancer should be less than 2 cm and there should not be node enlargement.

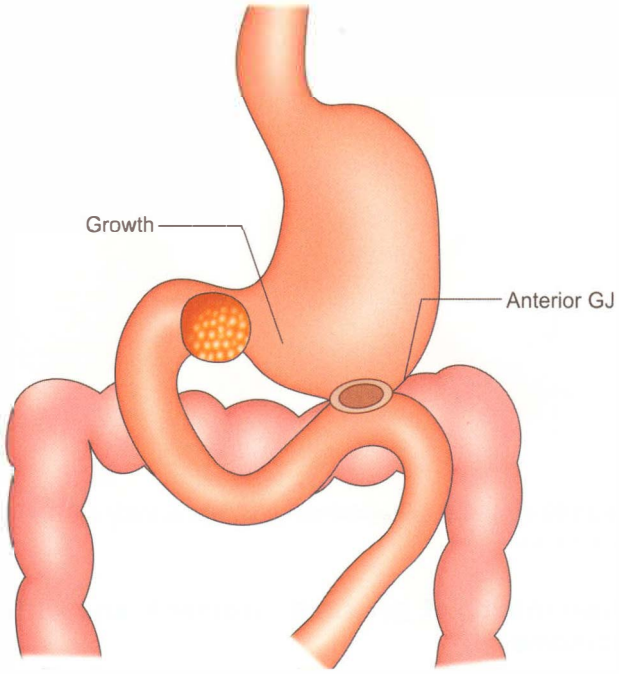


Fig. 23.96: Palliative anterior GJ

- Ideally cancer should be elevated variety and well differentiated.
- Normal saline is injected into submucosal plane and lesion gets elevated.
- It is excised with 1 cm margin up to muscularis propria at a deeper plane.

Endoluminal gastric surgery

- Small, high up lesions are ideal.
- Here, laparoscopic instrumentation is done under endoscopic guidance.
- Stomach is suitable for endoluminal surgery because it can be distended and contents are sterile.

KEY POINTS ON SURGERY FOR CARCINOMA STOMACH

1. Carcinoma antrum and body—radical subtotal gastrectomy
2. Carcinoma proximal stomach—radical total gastrectomy
3. Inoperable carcinoma distal stomach—palliative anterior GJ.
4. Radical refers to removal of lymph nodes, fat, fascia greater and lesser omentum.
5. When level 1 to 6 (N1) nodes are removed along with stomach, it is D1 gastrectomy
6. When level 7–11 (N2) lymph nodes are also removed along with 1–6 nodes and stomach, it is called D2 gastrectomy.
7. Early gastric cancer without lymph nodes can be treated with endoscopic mucosal resection or endoluminal gastric resection.
8. Palliative gastrectomy (subtotal) is worth considering in appropriate cases of obstruction/bleeding.

Paraneoplastic syndromes associated with carcinoma stomach

- *Trousseau's syndrome*—Thrombophlebitis
- *Acanthosis nigricans*—Hyperpigmentation of the axilla and groin.
- Peripheral neuropathy.

THE ADJUVANT TREATMENT

- Now it is understood that gastric cancers partially respond to chemotherapy—in about 30% of cases given at advanced stage (results are better than cancer colon). Injection 5-FU (fluorouracil) 500 mg IV daily for five days, every 28 days. It can be given by IV infusion or IV bolus over 15 minutes.
- **Mechanism of action:** It is an antimetabolite and acts by interfering with DNA synthesis. Side-effects are myelosuppression, mucositis, excessive lacrimation, nausea, vomiting, etc.
- Combination of 5-FU with adriamycin, mitomycin and cisplatin has also been tried. However, toxicity is more with these drugs. FAM (5-fluorouracil, adriamycin and mitomycin C) and ECF (epirubicin, cisplatin and 5-FU) are popular agents.
- Intraperitoneal mitomycin and mitomycin C—impregnated charcoal have also been used (target the recurrence site-gastric bed).

1. Postoperative chemotherapy: Depends on the type of resection done

A. R0 resection (free margins): Any tumour greater than T2 including selected T2 cases should be considered for adjuvant therapy that includes 1 cycle of 5-FU+/– leucovorin OR capecitabine chemotherapy ⟶ followed by 5-FU+/– leucovorin OR capecitabine based chemoradiation ⟶ 3–4 more cycles of 5-FU+/– leucovorin OR capecitabine chemotherapy.

B. R1 (microscopic margin positive) and R2 (grossly positive margins) regardless of the TMN stage should receive chemoradiation followed by further cycles of adjuvant chemotherapy.

2. If the patient has received preoperative chemotherapy/chemoradiation then:

A. Adjuvant treatment is usually recommended beyond T2N0M0 which is usually 5-FU+/– leucovorin OR capecitabine based.

B. If patient has received ONLY preoperative chemotherapy and the resection was R1/R2, then postoperative chemoradiation is advised.

Neoadjuvant chemoradiation

- It is advised in medically fit unresectable—locally advanced diseases which generally includes concurrent 5-FU/capecitabine-based chemotherapy.

- Chemotherapy alone may also be used as a neoadjuvant modality.
- Recommended radiotherapy dose is 45–50.4 Gy (at 1.8 Gy/fraction).

Metastatic gastric cancer:

- **Trastuzumab (Herceptin) can be used in metastatic or locally advanced gastric adenocarcinomas which are overexpressing HER-2 neu receptors (recent recommendation)**
- External beam radiotherapy can also be used in metastatic cases as a part of palliation, especially when there is tumour bleed, gastric outlet obstruction due to the tumour pain. Dose for palliative radiotherapy is generally 30 Gy in 10 fractions over 2 weeks.

CLINICAL NOTES

A 37-year-old female was admitted with vomiting, early satiety and abdominal pain of 20 days duration. Clinical examination revealed anaemia. No mass was palpable. UGI scopy revealed an ulcer in the lesser curvature and biopsy was reported as chronic gastritis—to rule out H. pylori infection. Ultrasound showed diffuse wall thickening (8–10 mm) in the anterior and posterior walls of the stomach. Barium follow revealed hour glass contracture probably secondary to chronic gastric ulcer. At surgery, wide stomach was thickened. Total gastrectomy (D-1) was done. Specimen when opened, revealed extensive infiltrative cancerous tissue. Final report was diffuse infiltrative adenocarcinoma. In a doubtful ulcer, investigate thoroughly and if necessary, do a laparoscopy/laparotomy.

OTHER TUMOURS OF THE STOMACH

GASTROINTESTINAL STROMAL TUMOURS (GISTs)

- Previously named leiomyoma, leiomyosarcoma is called GIST today.
- In gastrointestinal tract, **stomach is the commonest** site of GIST.
- **Bleeding** is the commonest presentation. It occurs due to ulceration of mucosa which on endoscopy gives **appearance of cervix** (Fig. 23.97).
- Larger tumours can present with dysphagia, weight loss, anaemia and palpable mass (Fig. 23.98).
- ***The diagnosis of malignancy is by greater mitotic figures.***
- **If mucosa is not ulcerated, 'biopsy on a biopsy'** or 'well biopsy' is necessary.
- Immunohistochemistry is mandatory for diagnosis—**C Kit gene amplification: It is the tumour marker of GIST**
- CT scan is a useful investigation (Figs 23.99 and 23.100).
- Resection is the best treatment (Figs 23.101 to 23.103).

Fig. 23.97: Bleeding ulcer of the stomach

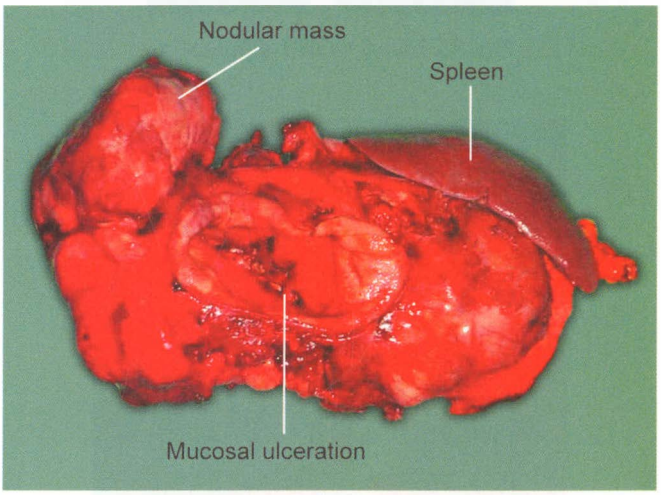


Fig. 23.98: A large GIST arising from the fundus of the stomach presenting as a nodular mass in the left hypochondrium resected successfully along with spleen

Fig. 23.99: CT GIST fundus

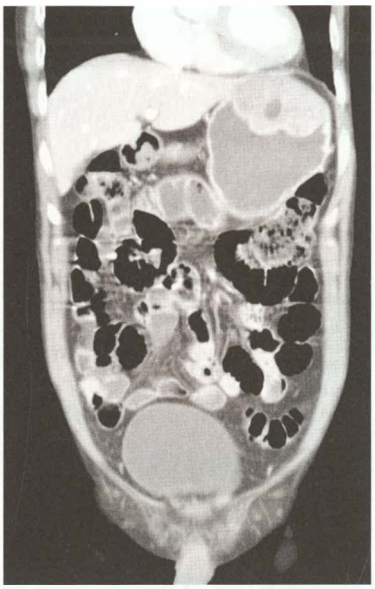


Fig. 23.100: CT GIST different cut

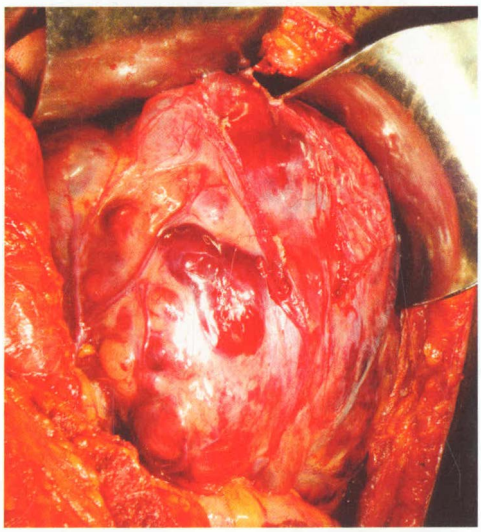


Fig. 23.101: GIST at surgery

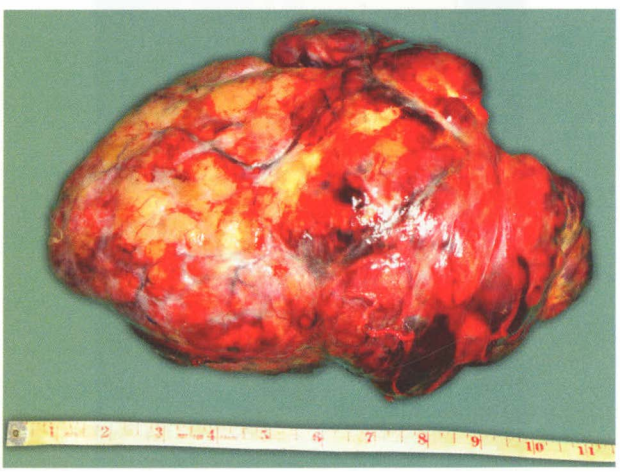


Fig. 23.102: GIST specimen

Fig. 23.103: Stomach opened to show fleshy mass

Chemotherapy for GIST

Capsule Imatinib 100 mg 4 capsules—to a total of 400 mg per day is given till toxicity appears or metastasis or residual tumour regresses (investigated by CT scan). This is a promising new drug.

GASTRIC LYMPHOMA

- Incidence of primary gastric lymphoma is increasing.
- They are B-cell derived from mucosa associated lymphoid tissue (**MALT**)—**MALTOMA**.
- Pain, weight loss, bleeding are common presentations.
- **6th decade** is the common age group.
- Endoscopic features are not specific but diffuse thickening with or without ulcerations may be seen.
- It is important to rule out **generalised process** by CT, ultrasound, bone marrow aspirate.
- Gastrectomy is the best treatment (Fig. 23.104).
- Chemotherapy is better for systemic disease.

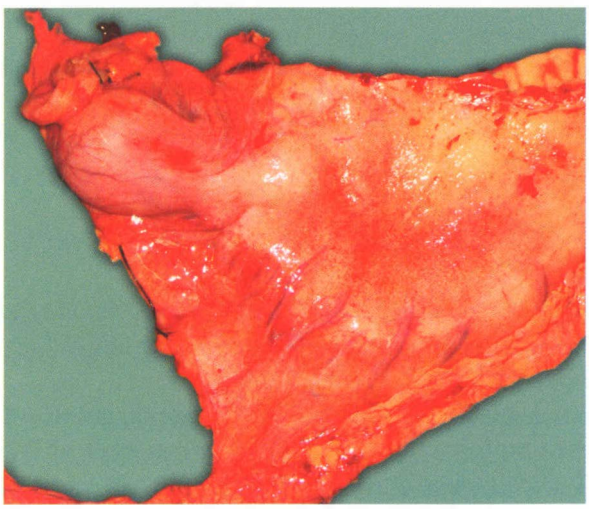


Fig. 23.104: Diffuse lymphoma—total gastrectomy specimen

PEARLS OF WISDOM

Low-grade lymphoma associated with H. pylori infection may regress and totally disappear after eradication treatment for H. pylori.

- Can be classified as complications of vagotomy, complications of GJ and complications of gastrectomy.

COMPLICATIONS OF VAGOTOMY

1. Stasis of food in the stomach resulting in nausea, loss of appetite, distension of upper abdomen, foul eructation, etc.
2. Denervation of gall bladder can cause gall stones.
3. Postvagotomy **diarrhoea**—can be very troublesome at times (Key Box 23.28).
4. Vagotomy produces hypoacidity which allows bacterial proliferation. Nitrates are reduced to nitrites which are carcinogenic. Such a malignancy which develops at GJ site is called **stump carcinoma**.

KEY BOX 23.28

CAUSES OF DIARRHOEA

- Vagotomy results in removal of parasympathetic influence on the function of foregut and midgut.
- Fast gastric emptying occurring due to GJ or gastrectomy.
- Hypoacidity resulting in bacterial proliferation causing enteritis.
- Bile salts also play a role.

COMPLICATIONS OF GJ

1. Stomal obstruction

It is due to oedema as in gastroduodenal anastomosis or nondependent drainage as in GJ. Sometimes, fat in the transverse mesocolon undergoes necrosis resulting in obstruction to the loops. Stomal obstruction also develops if there is narrowing of the lumen. Treatment is conservative. Surgery may also be required later, after confirming obstruction by gastrografin studies.

2. Retrograde jejunogastric intussusception (Fig. 23.105)

It develops if efferent and afferent loops are not sutured properly. It can appear at any time after surgery.

Clinical features

- Previous history of abdominal surgery (surgery done for peptic ulcer)
- Acute abdominal pain in upper abdomen
- Vomiting, sometimes blood stained
- **Palpable mass** in the upper abdomen.

Investigation

Barium meal X-ray shows filling defect in the stomach (Fig. 23.106). Sometimes, following a barium meal, intussusception is reduced (Fig. 23.107).

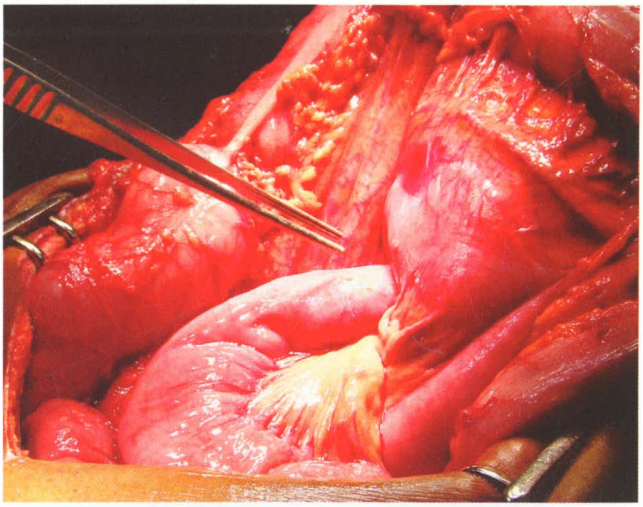


Fig. 23.105: Jejunogastric intussusception

Fig. 23.106: Barium showing filling defect, in a patient who had undergone GJ

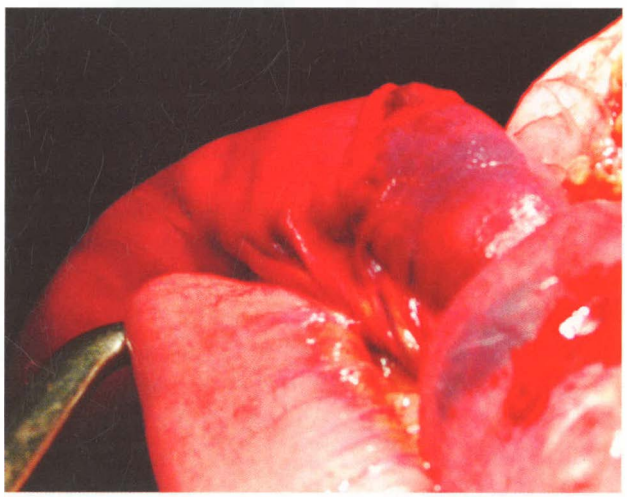


Fig. 23.107: Gangrene in jejunogastric intussusception

Treatment

Reduction of the intussusception and suturing of intestinal loops properly. If the loops are gangrenous, resection may be necessary (Figs 23.108 and 23.109).

Complication

Gangrene of intestine (Fig. 23.107 and 23.108).

Fig. 23.108: Gangrene of the intestines in a patient who had jejunogastric intussusception

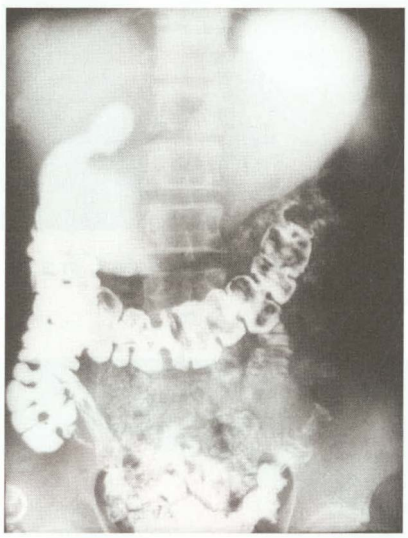


Fig. 23.109: Barium enema —barium entering into the stomach from colon (barium meal is not the investigation for gastrojejunocolic fistula)

3. Gastrojejunocolic fistula

- It is a complication of GJ done for peptic ulcer, especially when vagotomy is not done or is incomplete.
- After a few years of GJ, a recurrent ulcer can develop at the stoma—GJ site.
- This recurrent ulcer slowly invades the adjacent structure such as the transverse colon resulting in gastrojejunocolic fistula.

Clinical features

- Previous history of vagotomy and GJ
- Foul eructation and foul vomiting due to colonic contents entering the stomach which is loaded with faecal matter and foul contents.
- Intense diarrhoea due to severe jejunitis brought about by colonic bacteria entering the jejunum.
- Rapid deterioration in health—loss of weight, loss of appetite, dehydration and emaciation.

PEARLS OF WISDOM

Diarrhoea is not due to food entering the colon. Contents of the colon enter the stomach and then jejunum resulting in jejunitis causing diarrhoea.

Diagnosis

- Confirm by barium enema—barium entering the stomach (because of high pressure in the colon). Barium meal study should not be done (Fig. 23.109).

Treatment

- Triple resection (Figs 23.110 to 23.112)
- Preoperative preparation is necessary in the form of blood transfusion, stomach wash, nutritional supplementation and correction of dehydration.
- Resection of portions of stomach, intestine and colon followed by end to end anastomosis.

4. Stump carcinoma (Figs 23.113 to 23.115)

- It refers to carcinoma developing in the stomach after some surgery on the stomach. Classically it happens after a gastrojejunostomy (GJ) Billroth II or after a pyloroplasty.
- It develops after 10 to 20 years of surgery.
- Reflux of bile, changes in the acidity due to vagotomy are a few factors that precipitate stump carcinoma. Metaplasia is due to enterogastric reflux and bacterial reduction of nitrates.
- Clinical features include sudden loss of appetite, loss of weight with or without mass abdomen.
- Diagnosis is by endoscopy (Fig. 23.114).
- Treated by resection. However, many cases are advanced and they are inoperable.

GASTROILEOSTOMY (Fig. 23.116)

- It is an avoidable complication.
- Instead of the short loop—jejunum, ileum is anastomosed to the stomach.
- There will be severe uncontrolled diarrhoea, loss of weight and emaciation within a short period.
- Barium meal with fluoroscopy should be done which shows rapid flow of barium from stomach into the ileum.
- Laparotomy—undoing of gastroileostomy and fresh gastrojejunostomy should be done.

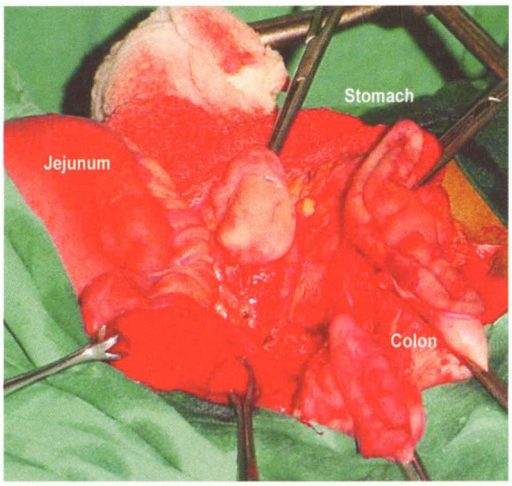


Fig. 23.110: Specimen of triple resection

Fig. 23.111: Gastrojejunocolic fistula —gastroscopy shows faecal matter in the stomach, coming from colon (*see* the triangular folds of colon)

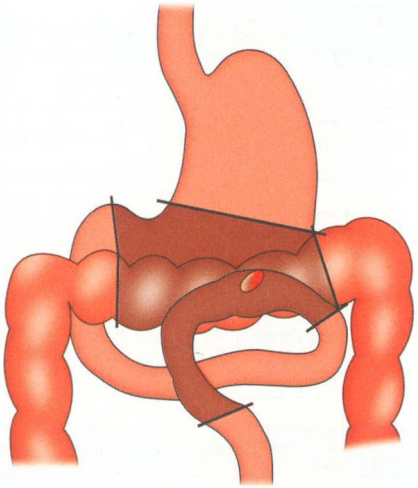


Fig. 23.112: Triple resection

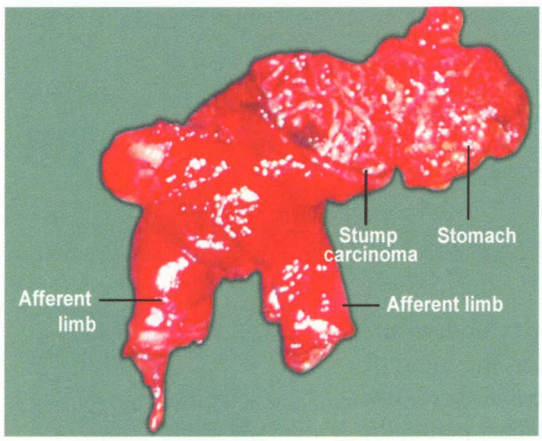


Fig. 23.113: Gastrectomy for stump carcinoma

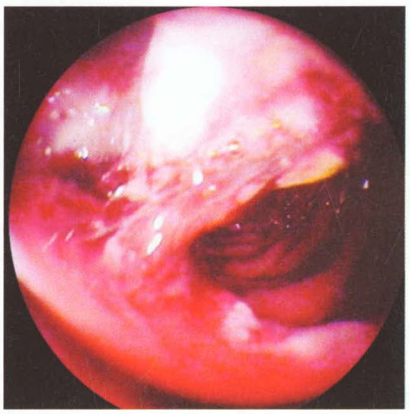


Fig. 23.114: Endoscopy showing stump carcinoma

Fig. 23.115: GJ was done in this patient 20 years back. He came with stump carcinoma which was inoperable

Fig. 23.116: Gastroileostomy—the biggest blunder a surgeon can do—it can be avoided by identifying the duodenojejunal flexure

POSTCIBAL SYNDROMES

This syndrome complex results due to rapid emptying of stomach contents to the distal intestines resulting in various physiological changes such as vasomotor symptoms, hypoglycaemia, etc. They are of two types. Their comparison is given in Table 23.10.

PEARLS OF WISDOM

Alkaline reflux gastritis which develops after GJ should be differentiated from afferent loop syndrome (*see* Table 23.11 on the next page).

COMPLICATIONS OF GASTRECTOMY

1. NUTRITIONAL DISTURBANCES

- Vitamin B_{12} and calcium deficiency.
- Megaloblastic anaemia occurs late due to gastric mucosal atrophy.
- **Iron deficiency anaemia**, common after gastrectomy when duodenum is bypassed because of deficient iron absorption.

Table 23.10 Dumping syndrome—postcibal syndromes

		Early	Late
1.	Onset	Increased emptying immediately after meals	One to two hours afterwards.
2.	Causes	Rapid passage of food of high osmolarity from stomach into small intestine causes hypovolaemia	Initial hyperglycaemia causes increased release of of insulin which is followed by hypoglycaemia.
3.	Relief	Lying down	Glucose
4.	Aggravating factors	More food, glucose, fullness	Exercise
5.	Symptoms	Epigastric distension, sweating, diarrhoea	Tremors, fainting attacks (release of adrenaline)
6.	Treatment	Small quantity, dry food. Over a period of time, symptoms settle down, less sugar content	Dietary adjustment of food

Table 23.11 Comparison of chronic afferent limb syndrome and alkaline reflux gastritis

	Chronic afferent limb syndrome	Alkaline reflux gastritis
• Pain	After meals	Unrelated to meals
• Vomitus	Bile	Bile and food
	Relieves pain	No changes in pain
	Projectile	Nonprojectile
• Occult bleeding	Rare	Common
• Cause	Limb obstruction—long loop	Enterogastric reflux (no obstruction)
• Treatment	Avoid a long loop at surgery	High fat, amino acid regimen
	Convert Billroth II to Billroth I	Proton pump inhibitors
	Roux-en-Y jejunostomy	Ursodeoxycholic acid
		Jejunojejunostomy, converting Billroth II to Billroth I can be done

- **Diarrhoea** is due to vagotomy causing intestinal hurry or due to dumping.
- Due to poor nutrition, there is **weight loss** and they are susceptible for **pulmonary tuberculosis**.

2. DUODENAL FISTULA (duodenal blow out)

It is the leakage of duodenal contents to the exterior. It commonly occurs after surgery.

Causes

- After a partial gastrectomy/total gastrectomy, where the closure of duodenum was difficult.
- After closure of perforated duodenal ulcer, which gives way once again.
- Injuries to duodenum during right hemicolectomy, right nephrectomy, etc.

Precipitating factors

- Faulty technique of closure of duodenal stump.
- Severely inflamed duodenum due to an active ulcer.
- If there is a distal obstruction, it increases tension in duodenal loop and may result in fistula.
- Ischaemia of duodenal stump.

Clinical features

- Signs and symptoms develop usually after 4 to 5 days when oral fluids are commenced. These stimulate outpouring of biliary and pancreatic juices.
- Severe upper abdominal pain and guarding, rigidity, hypotension and shock-like features of biliary peritonitis develop if there is no drainage tube.
- If drainage tube is kept in the first surgery, bile flows to the exterior. In such cases, signs of peritonitis are usually not present. However, severe electrolyte imbalance can occur (Fig. 23.117).

Fig. 23.117: Duodenal blow out following gastrectomy

Treatment

- Conservative treatment is successful in majority of the cases. Fistula heals in a few days, provided there is no distal obstruction. During this time hydration, electrolyte care is essential. Appropriate antibiotics are given.
- Surgical—if the fistula persists, laparotomy and closure of the fistula can be done by repairing with nonabsorbable sutures.

Complications

- Biliary peritonitis
- Septicaemia if bile is not drained outside.

- Excoriation of abdominal skin can be prevented by zinc oxide application.
- Fluid and electrolyte imbalance.

3. RECURRENT ULCER

It can be true anastomotic ulcer (gastrojejunal, gastroduodenal or jejunal ulcer), or a gastric ulcer in the remnant, or recurrent ulcer following highly selective vagotomy (HSV).

Incidence

- 3% after Billroth II gastrectomy
- 5 to 8% after vagotomy and GJ
- 40% after gastrojejunostomy
- 10 to 12% following HSV.

Causes of recurrent ulcer (Fig. 23.118)

1. Incomplete vagotomy
2. GJ alone
3. Inadequate gastrectomy
4. Narrow stoma
5. Zollinger-Ellison syndrome
6. Hyperparathyroidism

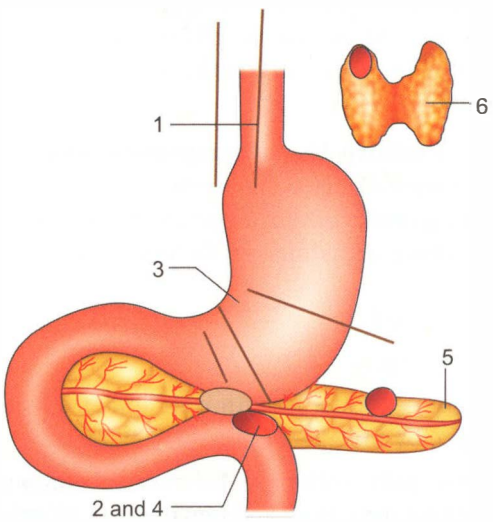


Fig. 23.118: Various causes of recurrent ulcer (*see* the text for numbers)

Symptoms

- Appears within 1 to 2 years after the operation.
- Severe persistent pain, 'boring' type, which gets worse within a few minutes of taking food. The pain is felt on the left side of the abdomen, near the umbilical region and it passes downwards.
- The pain is felt in the lower left chest following antecolic anastomosis. It is not relieved by antacids or milk unlike peptic ulcer. Bleeding may manifest as haematemesis, melaena or anaemia. Perforation can occur, resulting in peritonitis.

Diagnosis

- Gastroscopy gives the correct diagnosis.
- Hypercalcaemia and hypergastrinaemia should be ruled out.

Management (Table 23.12)

- Conservative treatment with H_2 receptor blockers is nearly always effective but relapse occurs if they are stopped. Smoking should be stopped.
- However, definitive surgery is indicated in appropriate cases.

ACID FUNCTION TESTS

These tests are not routinely done nowadays due to the availability of endoscopy facilities. However, in rare cases of recurrent peptic ulcer disease or as in Zollinger-Ellison syndrome, these tests are done. Hence, these are discussed in the last pages.

PENTAGASTRIN TEST

It is done to assess peak acid output.

Principle

Pentagastrin stimulates parietal cell mass resulting in outpouring of gastric acid.

Procedure

- Basal secretion of fasting stomach is measured.
- 6 mg/kg body weight of pentagastrin is administered subcutaneously or intramuscularly.

Table 23.12 Management of recurrent ulcer

Type of first surgery		Corrective surgery
1. GJ alone	→	Vagotomy
2. Vagotomy + GJ (incomplete vagotomy)	→	Incomplete vagotomy is the cause. Usually posterior vagus is found and it has to be divided
3. Vagotomy + GJ (complete vagotomy)	→	Stoma is not adequate. Partial gastrectomy is the ideal treatment
4. Billroth partial gastrectomy	→	Vagotomy with or without revision gastrectomy
5. HSV	→	Vagotomy + partial gastrectomy

- 4 samples of stomach secretion are collected for one hour, once every 15 minutes.
- By using suitable formula, peak acid output is measured.

Results

- Very high values are found in Zollinger-Ellison syndrome.
- Its values are very high in duodenal ulcer patients. Vagotomy and antrectomy may be the treatment of choice.
- It has a role in recurrent ulcer.
- In patients with gastrinoma, the basal acid output is unusually high and there may be a little response to pentagastrin stimulation.

HOLLANDER'S TEST (INSULIN TEST)[1]

- It is done to know the completeness of vagotomy.

Principles

- Insulin produces hypoglycaemia which stimulates vagus which in turn stimulates the parietal cell mass to secrete acid.
- If vagotomy is complete, there is no change in acid output during insulin test—Hollander's test.

Procedure

- Aspirate the fasting stomach contents
- To a fasting patient, 0.2 units /10 kg body weight of insulin is given subcutaneously.
- Blood sugar is estimated at 15-minute intervals and it is maintained between 30 and 40 mg% after two hours.

Results

Acid output is measured for one hour. If there is no change in acid output, vagotomy is complete. If there is a rise in concentration of 20 mmol per litre above the basal level in the first hour, it suggests incomplete vagotomy.

Usefulness

- In recurrent ulcers to know whether vagotomy is complete or not.
- To diagnose Zollinger-Ellison syndrome, where very high values of acid are seen.

Complications

Hypoglycaemia and coma. This test is obsolete now.

NIGHT FASTING SECRETION (DRAGSTED TEST)

The secretions of the stomach in the resting period or interdigestive period for 12 hours in the night are measured.

Procedure

Introduce a Ryle's tube and aspirate the stomach contents fo 12 hours from 9 PM to 9 AM. The volume and HCl in thi gastric juice are measured.

Results

- In normal patients, the total amount of gastric secretion i around 400 ml. Above this, it suggests vagal hyperactivity
- In Zollinger-Ellison syndrome the levels may be as high a one litre.
- Free HCl in normal patients is 10–20 mEq/L, in duodena ulcer 60–80 mEq/L, in gastric ulcer 10–20 mEq/L, and ir Zollinger-Ellison syndrome, it may be around 100–30(mEq/L.

ACUTE DILATATION OF STOMACH

Aetiopathogenesis

1. Can occur after any operation, particularly splenectomy and pelvic procedures.
2. It can occur following fracture femur, application of plaster of Paris, etc. Malnutrition, excessive distension of the stomach due to ventilation, aerophagia are other precipitating factors.

There is a **sudden loss of sympathetic tone** resulting in massive dilatation of stomach. Improper Ryle's tube aspiration and permitting intake of oral fluids too early before paralytic ileus settles down are additional factors.

Clinical features

- History of surgery
- **Hiccoughs**—due to irritation of under surface of the diaphragm, by the hugely distended stomach.
- Abdominal pain, vomiting, distension. Vomiting contains foul smelling dirty fluid and blood and is effortless.
- Features of shock. In untreated cases, can lead to cardiovascular collapse.
- **Effortless vomiting** of litres of dark watery fluid is characteristic of this condition.

Treatment: Urgent resuscitation

- **Introduce a Ryle's tube and aspirate the stomach. It is the life-saving use of Ryle's tube (Fig. 23.119).**
- Rapid IV fluid replacement, with normal saline and dextrose saline. Both crystalloids and colloids may be necessary to treat the shock and electrolyte abnormalities.

[1]Many deaths have occurred due to this test in the past.

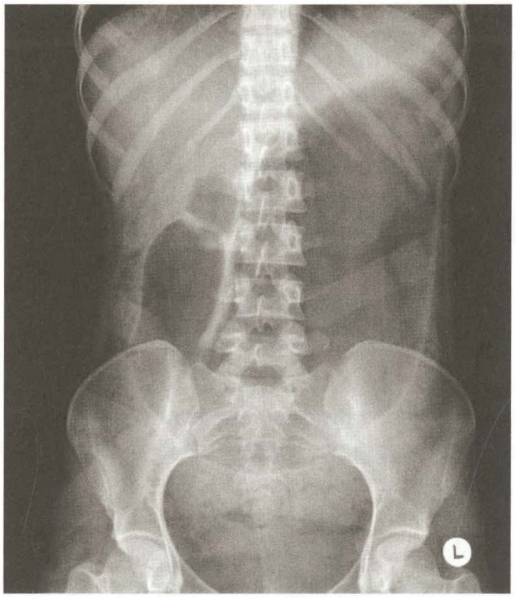


Fig. 23.119: Plain X-ray showing acute dilatation of the stomach. Ryle's tube insertion is a life-saving simple procedure in this condition

Complications

- Pulmonary: In debilitated patients, aspiration may result in aspiration pneumonitis (Mendelson syndrome).
- It carries significant mortality.

VOLVULUS OF THE STOMACH

- It is a rare condition in which stomach rotates in a horizontal (organoaxial) and vertical (mesentericoaxial) direction resulting in an acute abdomen.
- Organoaxial is more common.
- Many times, volvulus is intermittent.
- In general, initially the colon moves upwards and later greater curvature of the stomach.
- There is associated eventration of the diaphragm which also precipitates this condition (in children, congenital).
- In adults, diaphragmatic defects are more commonly traumatic or paraoesophageal herniation.
- Clinical features include epigastric pain, fullness and tenderness.

Borchardt's triad

1. Sudden, constant, severe upper abdominal pain
2. Recurrent retching with production of little vomitus
3. Inability to pass a nasogastric tube.

Diagnosis

- Plain X-ray abdomen/chest: Gas filled viscus
- Barium meal can demonstrate twisted stomach
- Upper GI scopy.

Treatment

- Reduce the volvulus by dividing gastrocolic omentum
- Fix the greater curvature of the stomach to the duodeno-jejunal flexure or perform a GJ without stoma.
- Repair of eventration
- Fixing by tube gastrostomy can also be done.

BEZOARS

- Bezoars are collections of **nondigestible materials,** usually of vegetable origin (phytobezoar) but also of hair (trichobezoar).
- **Trichobezoars** are concentrations of hair, generally found in long-haired girls or women who often deny eating their own hair (trichophagy) (Figs 23.120 and 23.121).

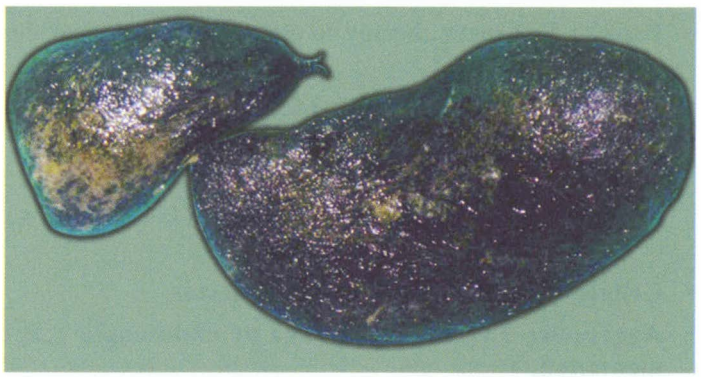


Fig. 23.120: Trichobezoar: Appreciate the shape of stomach and first part of the duodenum in bezoars (*Courtesy:* Dr Rajiv Shetty, Prof & Head, Dept of Surgery, Bangalore Medical College and Medical Superintendent, Bowring Hospital, Karnataka)

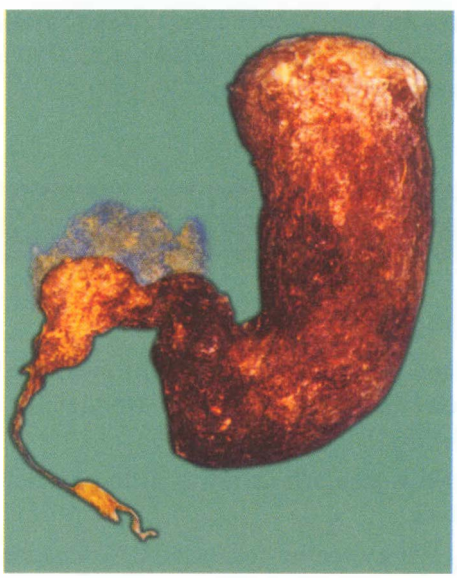


Fig. 23.121: Trichobezoar (*Courtesy:* Dr YV Krishna Rao, Prof of Surgery, Mamata Medical College and Hospital, Khammam, Andhra Pradesh)

Causes

- **Phytobezoars** are most commonly found in patients who have undergone surgery of the stomach and have impaired gastric emptying.
- **Diabetics with autonomic neuropathy** patients are also at risk.

Clinical features and diagnosis

- The symptom of gastric bezoars include early satiety, weight loss, nausea, pain and vomiting due to **gastric outlet obstruction** with occasional gastric perforation and small bowel obstruction.
- **A large mass** may be palpable on physical examination and the diagnosis is confirmed by a barium examination or endoscopy.

Treatment

- **Papain:** Enzymatic therapy to attempt dissolution of the bezoar.
  - Papain found in Adolph's Meat Tenderizer (AMT) is given in a dose of one teaspoon in 150 to 300 ml water several times daily.
  - The sodium concentration in AMT is high, so hypernatraemia may occur.
- **Cellulase** has been used with some success.
- **Aggressive Ewald tube lavage or endoscopic fragmentation.**
- Failure of these therapies would necessitate surgical removal.

PEARLS OF WISDOM

Trichobezoars tend to form a cast of the stomach, with strands of hair having been observed as far distally as the transverse colon.

IDIOPATHIC HYPERTROPHIC PYLORIC STENOSIS (IHPS)

Aetiopathogenesis

- In this condition there is **hypertrophy** involving the pyloric antral **circular muscle fibres**. Duodenum is normal (Fig. 23.123).
- The lumen is so much narrowed as to give rise to pyloric obstruction.
- Familial history can be obtained in a few patients.
- Prenatal incoordination of muscles, lack of nitric oxide relaxation of smooth muscles may be some factors responsible for IHPS.

Clinical features

- **Incidence:** 3–5/1000 births. First born male child is affected very often. The child is normal at birth and the symptoms appear around 6–8 weeks.
- First symptom is **vomiting**. It is **projectile**, forcible, does not contain bile. A visible gastric peristalsis (VGP) can be seen especially when the mother feeds the child (Fig. 23.122).
- Loss of weight—dehydration.
- Constipation and oliguria are the features.
- *Per* abdomen—hypertrophied thickened pylorus can be felt as a mass in the right hypochondrium—'Olive like mass' (in adults with pyloric stenosis due to chronic duodenal ulcer, no mass is felt—Fig. 23.123).

Ultrasound

- Long axis view of pylorus demonstrates 'cervix' sign
- Short axis view of pylorus demonstrates 'target' sign

Fig. 23.122: Visible gastric peristalsis

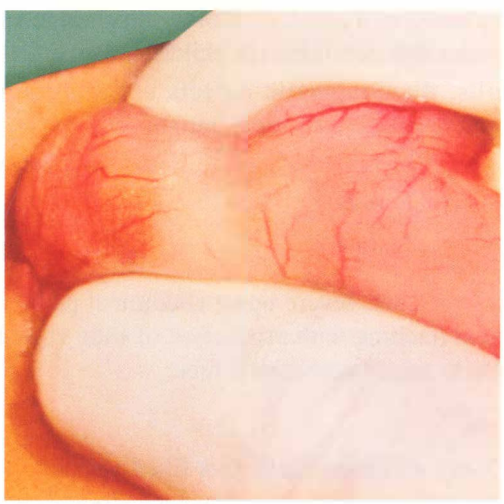


Fig. 23.123: Hypertrophied pylorus

- When muscle thickness is more than 7–15 mm size and pyloric canal length is 20–25 mm, it is diagnostic.

Treatment

- Correction of dehydration and electrolyte disturbance by intravenous half normal saline must always precede surgery.
- **Ramstedt's pyloromyotomy** is the surgical treatment (Figs 23.124 to 23.127).

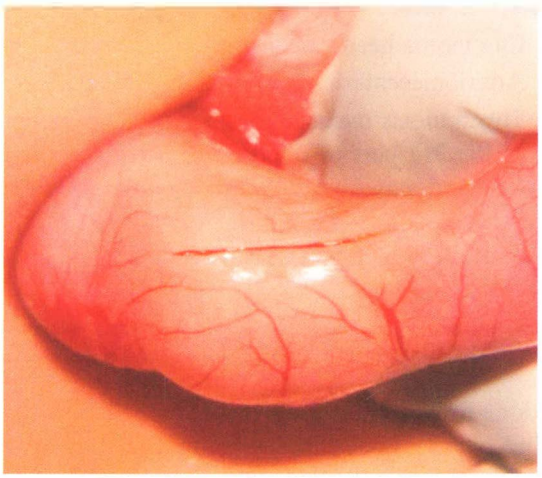


Fig. 23.124: Pyloromyotomy incision

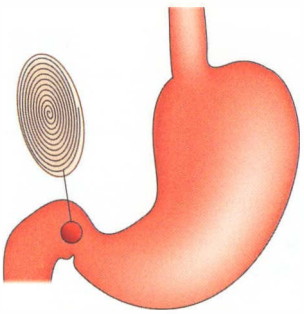


Fig. 23.125: Pyloric mass

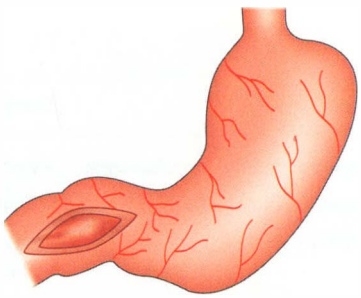


Fig. 23.126: Ramstedt's pyloromyotomy

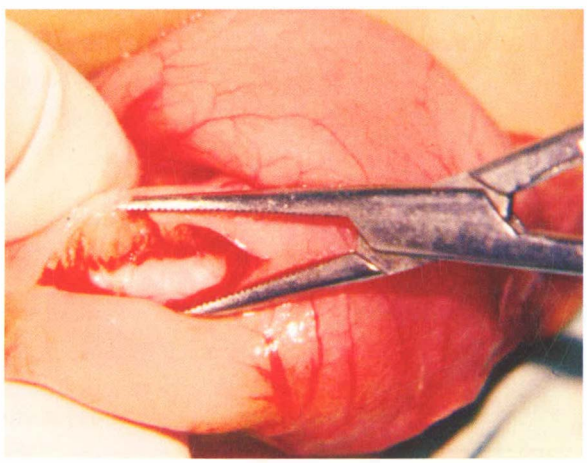


Fig. 23.127: Ramstedt's pyloromyotomy being done (*Courtesy:* Prof Vijaykumar, HOD of Paediatric Surgery, KMC, Manipal)

- Laparotomy is done and an incision is made through the serosa. It then cuts through the circular muscle fibres till all the muscle fibres are divided and the mucosa bulges out.
- Avoid injury to the mucosa. If mucosa is opened, it is sutured and reinforced by using omentum.

CHRONIC DUODENAL ILEUS

- It is also called superior mesenteric artery (SMA) syndrome, **Wilkie's disease or arteriomesenteric compression** (Figs 23.128 and 23.129). The third part of the duodenum is compressed between vertebral column and superior mesenteric vessels. Acute loss of fat and immobilisation with cast precipitates this disease. Hence, it is also called cast syndrome. Normal angle between superior mesenteric artery and aorta is 38–56°. Due to loss of fat or due to abnormal origin of the SMA, when the angle reduces to 6–25°, SMA syndrome occur (for risk factor—*see* Key Box 23.29).

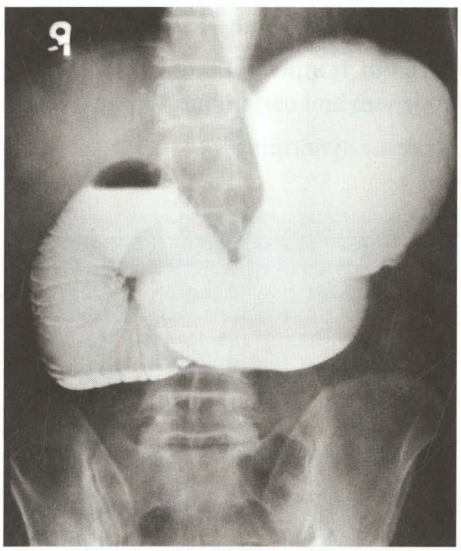


Fig. 23.128: Duodenal obstruction resulting in gross dilatation of duodenum—need for duodenojejunostomy than gastrojejunostomy in these cases

KEY BOX 23.29

RISK FACTORS FOR SMA SYNDROME

- **A**sthenic and tall—thin and tall body build—aortomesenteric angle is narrow (congenital)
- **B**urns—prolonged bed rest → loss of weight
- **C**achexia, catabolic phases, cast application
- **D**uodenal insertion is high at ligament of Treitz
- **E**maciation, malnutrition
- **F**emales twice as susceptible
- **G**ut malrotation/poor motility of the gut
- **H**yperextension of spine. Lumbar lordosis

ABCDEFGH of SMA syndrome

- In this condition, duodenum is dilated up to the 3rd part with obstruction to the 3rd or 4th part of duodenum (Fig. 23.128).
- Patients present with features of gastric outlet obstruction but vomitus contains bile—'Food Fear'.
- Abdominal pain is not severe as in intestinal obstruction but on careful questioning history of true nature of spasmodic pain is obtained.
- This condition is often misdiagnosed.
- Barium meal demonstrates the obstruction—dilated stomach and duodenum.
- Duodenojejunostomy is the treatment of choice (Fig. 23.130).

Differential diagnosis

1. **Pyloric stenosis:** In this condition, vomitus does not contain bile. Barium meal X-ray shows distended stomach. Duodenum is not seen.
2. **Annular pancreas:** Rarely, it can present in adults with obstruction to the second part of the duodenum. Vomitus may contain bile. Barium meal X-ray shows dilatation of the first part and also the second part of the duodenum.
3. **Tuberculosis of lymph nodes** can compress the 3rd part of the duodenum and can mimic Wilkie's disease.

What is nutcracker syndrome?[1]

DUODENAL ANATOMY AND OBSTRUCTION

- There are many causes of duodenal obstruction in children
- In adults important causes are given in Table 23.13.

1st part: Postbulbar ulcer with stenosis.

2nd part: 2 causes:

1. Annular pancreas in adults
2. Carcinoma head of pancreas

3rd part: 3 causes:

1. Carcinoma head of pancreas
2. Arteriomesenteric compression
3. Tuberculosis/metastatic nodes causing compression of the 3rd part of duodenum. Most common cause of duodenal obstruction in adults is carcinoma pancreas.

4th part: Carcinoma pancreas (Fig. 23.131)

GASTRIC SURGERY FOR MORBID OBESITY

- It is done for morbid obesity.
- Morbid obesity is defined as being 100% over the ideal weight for height or having body mass index (weight in kg/height in m^2) greater than 45.

Table 23.13 Anatomy of duodenum and surgical importance

Parts	Development	Length	Relationships	Blood supply	Surgical importance
1st part	• Foregut	2 inches	• Gall bladder • Liver • Bile duct • Portal vein	• Superior pancreaticoduodenal • Supraduodenal artery of Wilkie	• Perforation of 1st part (causes peritonitis) • Injury to duodenum in laparoscopic cholecystectomy
2nd part	• Foregut above ampulla of Vater • Midgut below ampulla of Vater	3 inches	• Right lobe of liver transverse colon anteriorly and kidney, renal vessels posteriorly		• Perforation of 2nd part manifests late because it is retroperitoneal • Injury to duodenum in right hemicolectomy
3rd part	• Midgut	4 inches	• Retroperitoneal fixed • Anteriorly crossed by superior mesenteric vessels • Posteriorly related to inferior vena cava	• Inferior pancreaticoduodenal artery	• Careful separation of mesenteric vessels from duodenum during pancreaticoduodenectomy
4th part	• Midgut	1 inch	• Anteriorly by transverse colon • Superiorly body of pancreas	• Inferior pancreaticoduodenal artery	• 4th part continues as duodenojejunal (DJ) flexure. It is an important landmark to identify first loop of jejunum during gastrojejunostomy

[1]Nutcracker syndrome is the entrapment of right renal vein between abdominal aorta and the SMA

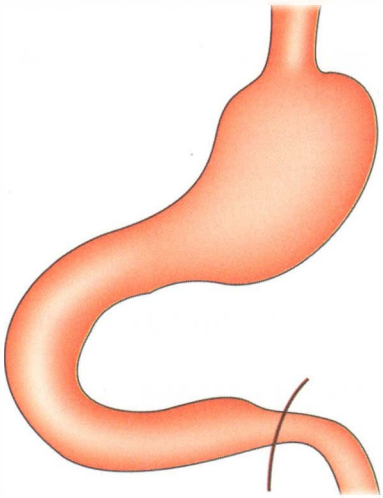


Fig. 23.129: Wilkie's disease—superior mesenteric artery syndrome

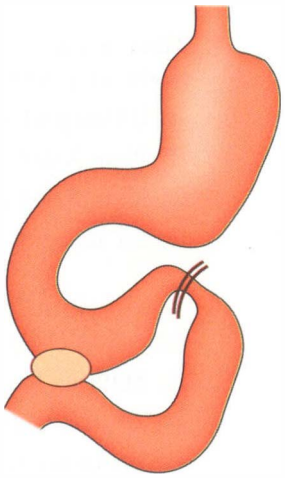


Fig. 23.130: Duodenojejunostomy

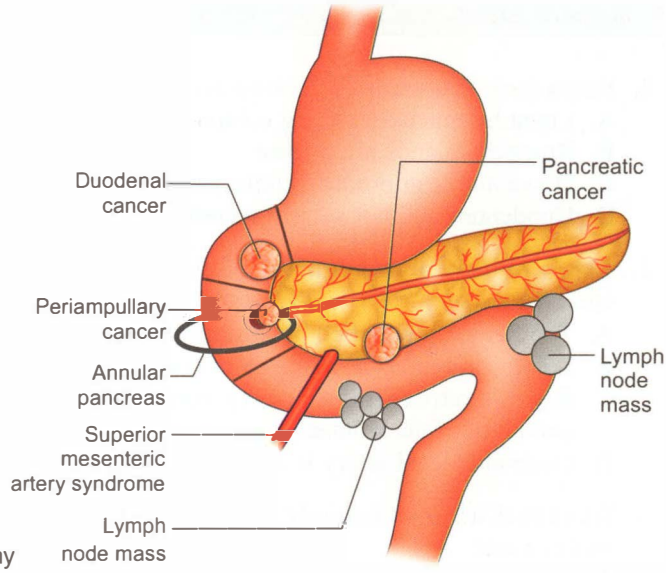


Fig. 23.131: Causes of duodenal obstruction

- Often these patients are young with diabetes and renal problems. Obesity related comorbidities such as diabetes mellitus, hypertension, coronary artery disease, osteo-arthritis, sleep apnoea, etc., will improve after surgery.
- Laparoscopic (lap) surgery is the choice.

1. **Lap band procedure:** It is one of the commonly performed bariatric procedure. In this procedure an inflatable cuff is placed laparoscopically **just below oesophagogastric junction**. It is adjustable. The balloon can be filled to 15 cc with saline. This can be increased or decreased also. Band slippage and stricture are problems. Band can also be removed. Advantage of banding is high safety and easy reversibility.
2. **Laparoscopic gastric bypass** is the procedure of choice with good results (Fig. 23.132).

Roux-en-Y gastric bypass (RYGB)

- 30 cc stomach pouch is fashioned using endostaplers.
- Duodenojejunal junction is identified by ligament of Treitz and jejunum is divided 100 cm distally.
- Distal loop is anastomosed to gastric pouch as GJ.
- Distal jejunojejunostomy is made at a distance of 150 cm.
- There is significant improvement in diabetes mellitus and hypertension, high cholesterol after weight loss surgery. A gastric pouch is created by using circular and linear staples.

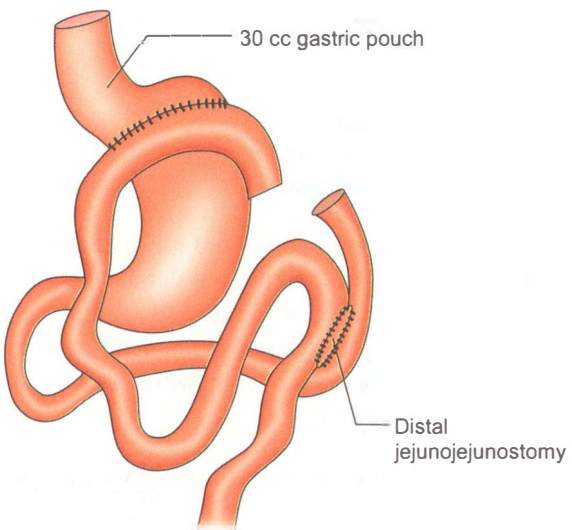


Fig. 23.132: Gastric bypass for morbid obesity

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been updated.
- New photographs and key boxes have been added.
- *Helicobacter pylori* infections and carcinoma stomach have been discussed in detail.
- Bezoars, surgery for morbid obesity have been added.
- Stump carcinoma, SMA syndrome have been explained in more detail.
- Chemotherapy for carcinoma stomach has been edited.

MULTIPLE CHOICE QUESTIONS

1. Regarding duodenum following are true *except:*
A. Lined by mucus-secreting columnar epithelium
B. Brunner's glands are present
C. Duodenum can produce cholecystokinin
D. Duodenum will not secrete secretin

2. Following are true regarding blood supply of the stomach *except:*
A. The fundus is supplied by left gastric artery
B. Left gastric artery is a branch of coeliac axis
C. Right gastroepiploic artery runs along greater curvature of the stomach
D. Gastroduodenal artery is a branch of hepatic artery.

3. Duodenal ulcer commonly occurs in which part of duodenum?
A. First part
B. Second part
C. Third part
D. Fourth part

4. Following are true for *Helicobacter pylori* bacteria *except:*
A. It is spiral shaped
B. It hydrolyses urea
C. It can produce cytotoxins
D. Mostly, it is associated with type A gastritis

5. Following are true for duodenal ulcer *except:*
A. It occurs in the first part of the duodenum
B. It may be associated with type B gastritis
C. Smoking may precipitate development of ulcer
D. It can erode branches of left gastric artery and result in massive bleeding

6. Following are true for malignancy in a gastric ulcer *except:*
A. It occurs rarely
B. Occurs in the lesser curvature of the stomach
C. It produces a notch in barium studies
D. Converging rugosity is lost

7. Most important cause of duodenal stump leakage after Billroth II gastrectomy is:
A. Efferent loop obstruction
B. Oedema of the stump
C. Kinking of the afferent loop
D. Fat necrosis

8. About truncal vagotomy which one of the following is not correct?
A. Decreases acid secretion
B. Decreases parasympathetic activity
C. Increases gall stones
D. Increases constipation

9. More accurate investigation for the diagnosis of the perforated duodenal ulcer is:
A. Ultrasound of the abdomen
B. Plain X-ray abdomen erect position
C. Plain X-ray chest
D. CT imaging

10. Treatment of the perforated duodenal ulcer include following *except*:
A. Suturing the ulcer in transverse direction
B. Truncal vagotomy
C. Peritoneal toilet
D. Omental patch

11. The most common site of bleeding from peptic ulcer is:
A. Lesser curvature of the stomach
B. Greater curvature of the stomach
C. First part of the duodenum
D. Fundus of the stomach

12. Following are true with Dieulafoy's lesion *except:*
A. It is an arteriovenous malformation
B. It is one of the causes of occult bleeding
C. Local excision is the choice of treatment
D. The site is fundus of the stomach

13. Which one of the following is the complication of gastrojejunostomy?
A. Gall stones
B. Duodenal blow out
C. Intussusception
D. Diarrhoea

14. Following are complications of vagotomy *except:*
A. Diarrhoea
B. Gall stones
C. Achlorhydria
D. Carcinoma stomach

15. Hypochloraemic alkalosis in pyloric stenosis is treated by:
A. Normal saline
B. Ringer lactate
C. Isotonic saline with potassium
D. Double strength saline

16. *H. pylori* is an important factor for carcinoma stomach in which region?

A. Distal
B. Proximal
C. Body
D. Gastro-oesophageal junction

17. Which one of the following is not a risk factor for proximal carcinoma stomach?

A. Chronic gastric ulcer
B. *H. pylori* induced gastritis
C. Pernicious anaemia
D. Obesity

18. Which one of the following is not a feature of early gastric cancer?

A. Confined to mucosa
B. Confined to mucosa and submucosa
C. Confined to mucosa with lymph nodes
D. Involving muscularis

19. Following are features of gastrointestinal stromal tumours (GISTs) of the stomach *except:*

A. Stomach is the most common site of GIST
B. More than 5 cm size tumour is considered to have malignant potential
C. Wedge excision is the treatment of choice for small lesions
D. Always do lymphadenectomy with wedge excision

20. Which one of the following features is false for gastrointestinal stromal tumour (GIST)?

A. It is arising from mesenchymal tissue
B. Associated with tyrosine kinase C-kit oncogene
C. They are sensitive to tyrosine kinase antagonist imatinib
D. Lymph node spread is very common

ANSWERS

1 D	2 A	3 A	4 D	5 D	6 C	7 C	8 D	9 D	10 B
11 C	12 D	13 C	14 D	15 C	16 A	17 B	18 D	19 D	20 D

24

Liver

- Surgical anatomy
- Physiology
- Pyogenic abscess
- Amoebic abscess
- Hydatid cyst
- Other cystic diseases
- Benign tumours
- Hepatoma
- Secondaries in the liver
- Portal hypertension
- Portal gastropathy
- Portal bliopathy
- Ascites in portal hypertension
- Budd-Chiari syndrome
- Role of octreotide in surgery
- Liver transplantation
- Haemobilia
- What is new?/Recent advances

SURGICAL ANATOMY OF THE LIVER

- Liver is located in the right hypochondrium extending into epigastrium and left hypochondrium.
- It weighs about 1,500 g—three quarters of it is the right lobe and rest is the left lobe (Figs 24.1 and 24.2).

Ligaments

1. **Right triangular ligament** fixes the right lobe to the undersurface of the right dome of diaphragm. When this ligament is divided, right lobe can be mobilised and can be turned to left as during right hepatic lobectomy or to control bleeding from liver trauma.
2. **Left triangular ligament** fixes the left lobe to the diaphragm in a similar fashion. For a complete vagotomy, when this ligament is divided, left lobe can be easily retracted—vagotomy becomes easy. This ligament also needs to be divided to get access to retrohepatic inferior vena cava.
3. **Falciform ligament:** It is the remnant of umbilical vein. It runs from umbilicus to the liver dividing the liver into right and left lobes, passing into the interlobar fissure. Division of superior leaves of falciform ligament helps in exposing suprahepatic inferior vena cava (IVC).

Blood supply

- 80% is derived from portal vein, 20% from hepatic artery.
- Right branch of hepatic artery supplies entire right lobe and left branch supplies the left lobe.
- Ligation of hepatic artery may not cause necrosis of liver in normal individuals. However, it can cause liver failure in cirrhotic patients (it was done in olden days to control secondaries in the liver), and in obstructive jaundice patients.

Venous drainage of the liver

- Major venous drainage is through 3 hepatic veins: right middle and left. They join the inferior vena cava (IVC) immediately below the diaphragm. *Right hepatic vein has a small extrahepatic course*. On the other hand, middle and left hepatic veins usually join within liver parenchyma.
- In fact, these three veins keep the liver suspended.

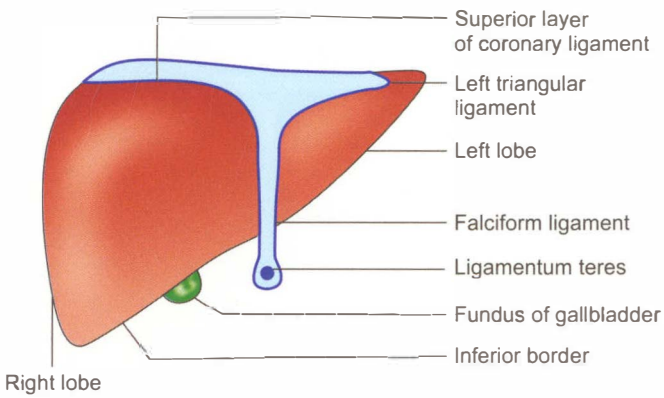


Fig. 24.1: Liver seen from the front

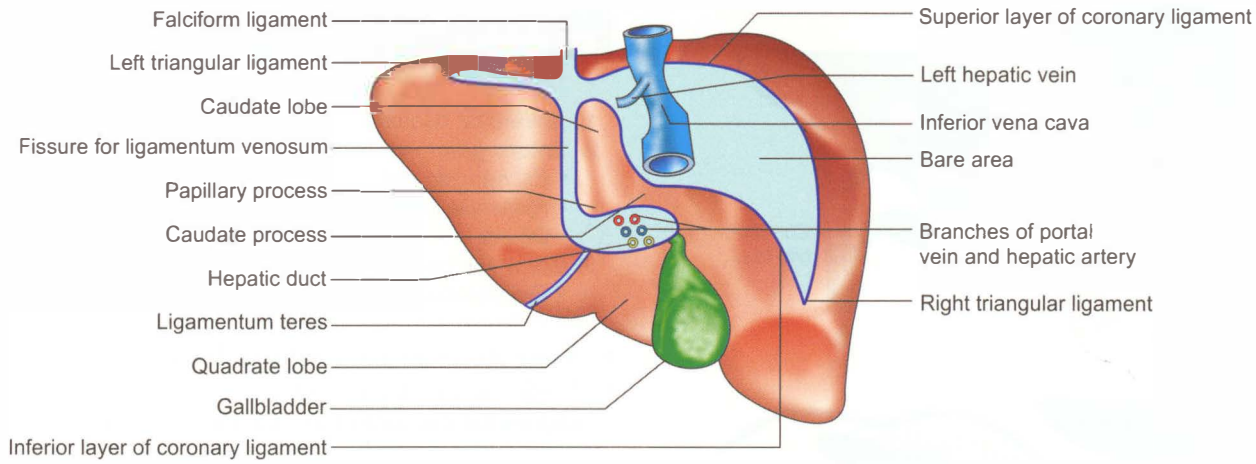


Fig. 24.2: Liver seen from behind and below

Lymphatics

- Superficial lobules: Subcapsular course to diaphragm or to suspensory ligament or posterior mediastinum.
- Deep lobules: Along IVC or portal vein into porta hepatis.

Hilum of the liver

- This is also called **porta hepatis**. It has the most vital structures within it—hepatic artery, bile duct anteriorly and portal vein posteriorly.
- These structures are present within the free edge of lesser omentum—hepatoduodenal ligament.
- Within the hilum, these structures divide into right and left branches. Left bile duct has a **longer extrahepatic course** of approximately 2 cm. In CBD strictures, this can be used for **hepaticojejunostomy**.

Regeneration

- Liver has an excellent **capacity for regeneration** after partial resection.
- A factor released from **pancreas** is supposed to be responsible for this.

Hepatic lobules

The ***functional unit*** of the liver is the hepatic lobule. Each hepatic lobule consist of sheets of liver cells, separated by hepatic sinusoids, venous channels which drain blood to central vein, a tributary of the hepatic vein from portal tracts that contain branches of the hepatic vein and portal vein.

Physiology (Table 24.1)

PYOGENIC LIVER ABSCESS

Causes (Fig. 24.3)

I. Infection through the portal vein
- Acute appendicitis
- Acute diverticulitis (sigmoid)
- Acute amoebic colitis
- Acute bacillary dysentery
- Ulcerative colitis

II. Infection through the common bile duct (CBD)
- Stricture of the CBD
- Periampullary carcinoma resulting in stasis of the bile, precipitating infection (cholangitis)

Table 24.1 Physiology of the liver

Functioning of the liver	Significance
• Principal site of synthesis of all proteins **except globulins**, which are produced in reticuloendothelial systems	• Hypoalbuminaemia in chronic liver disease
• **Glucose metabolism, glycolysis and gluconeogenesis**	• Persistent hypoglycaemia is a feature of fulminant hepatic failure (FHF), also seen in hepatoma
• Synthesis of **clotting factors**	• Bleeding tendencies in chronic liver disease
• Maintaining **core body temperature**	• Hypothermia in FHF
• Bilirubin formation	• Congenital hyperbilirubinaemia and other
• Drug and hormone metabolism	• Hepatitis due to drugs such as rifampicin and other antituberculous drugs
• Monitoring pH and correcting **lactic acidosis**	• Severely impaired in FHF
• Removal of endotoxins and foreign antigens	• Severely impaired in FHF
• Urea formation from protein catabolism	• After portocaval shunt, endotoxins bypass the liver, results in encephalopathy

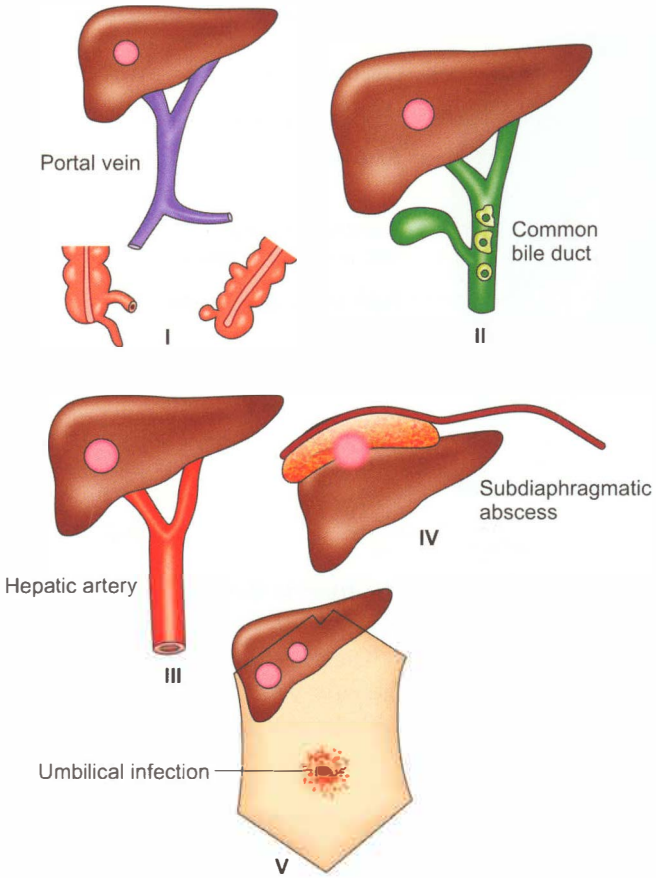


Fig. 24.3: Sources of pyogenic liver abscess

- Recurrent cholangitis due to stone in the CBD
- ERCP

III. Infection through the hepatic artery

- Septicaemia and pyaemia
- Actinomycosis of faciocervical region

IV. Extension abscess

- Subdiaphragmatic abscess
- Empyema thoracis
- Penetrating injuries

V. Infection through umbilicus

Neonatal umbilical sepsis giving rise to pyaemia.

Certain facts

- Majority of the infective bacteria are derived from gastro-intestinal tract (Key Box 24.1)
- In majority of cases, it is **polymicrobial** infection
- ***E.coli*** is the most common facultative organism
- ***Bacteroides fragilis*** is the most common anaerobe
- ***Candidal infection*** is increasing due to chemotherapy, especially for leukaemic patients.

Clinical features

- Alcoholic males and debilitated men suffer more, probably because of poor immunity.

KEY BOX 24.1

BACTERIOLOGY

1. Anaerobic bacteria	: 60% (*Bacteroides fragilis*)
2. Enteric gram –ve bacteria	
• *Escherichia coli*	: 40%
• *Klebsiella pneumoniae*	: 10–20%
• Others	: 4–40%
3. Gram +ve bacteria	
• *Staphylococcus aureus*	: 4–25%

- Acute abscesses are usually multiple, chronic are single.
- Tender hepatomegaly, low grade or high grade pyrexia with abdominal discomfort are the main features.

Investigations

- **Total WBC count** is raised.
- **Stool routine examination:** Amoebic cysts, culture and sensitivity for typhoid bacilli.
- **Abdominal ultrasound** and ultrasound-guided aspiration establishes the diagnosis.
- When in doubt, CT scan can be done (Figs 24.4 and 24.5), followed by FNAC which draws frank pus. Pus is sent for Gram's stain, culture and sensitivity. CT also helps in the diagnosis of ***associated conditions such as diverticulitis of the colon.***

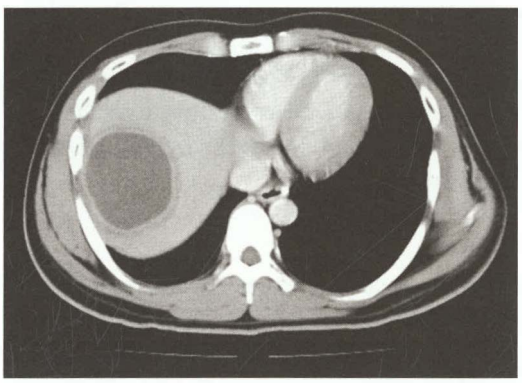


Fig. 24.4: CT scan showing hypodense lesion in the right lobe of liver suggestive of pyogenic liver abscess

Fig. 24.5: CT scan showing multiple liver abscesses proved to be due to amoebiasis

- Further investigations are directed towards the associated conditions. **Examples:**
  - **Chest X-ray:** Air under the diaphragm (perforation of hollow viscus) or diagnosis of empyema thoracis.

Treatment

1. **Conservative:** Multiple small abscesses may respond to antibiotics. However, they have to be given for 4 to 6 weeks.
2. **Percutaneous drainage** (Key Box 24.2)
   Method
   - Ultrasound or CT-guided aspiration and drainage by using a pigtail catheter.
   - Irrigation of abscess cavity with saline.
3. **Open (surgical) method** (Key Box 24.3)
   Laparotomy is required mainly to treat the primary causes, e.g. appendicectomy, drainage of appendicular abscess. If liver shows a significant abscess, it is drained. Alternately, a pigtail catheter is introduced into the abscess cavity and brought outside through a separate opening. It helps to drain for a longer period of time.
4. Laparoscopic drainage can also be done.

KEY BOX 24.2

PERCUTANEOUS DRAINAGE—INDICATIONS

- Superficial abscesses
- Abscess with no intra-abdominal pathology
- Abscess of unknown aetiology

KEY BOX 24.3

OPEN (SURGICAL) METHOD—INDICATIONS

- Abscess with intra-abdominal pathology
- Ascites
- Deep-seated abscess
- Multiple abscesses

AMOEBIC LIVER ABSCESS

It is also called **Tropical abscess** (dysenteric abscess). It is the commonest extraintestinal manifestation of amoebiasis.

Aetiopathogenesis

- This disease is caused by ***Entamoeba histolytica.***
- It is almost always a complication of amoebic dysentery. This can occur in the acute stage or in the chronic carrier stage.
  - **Infection from the caecum** (typhlitis) spreads through the tributary of **superior mesenteric vein.**
  - **From sigmoid colon**, through the tributary of **inferior mesenteric vein** (Fig. 24.6).

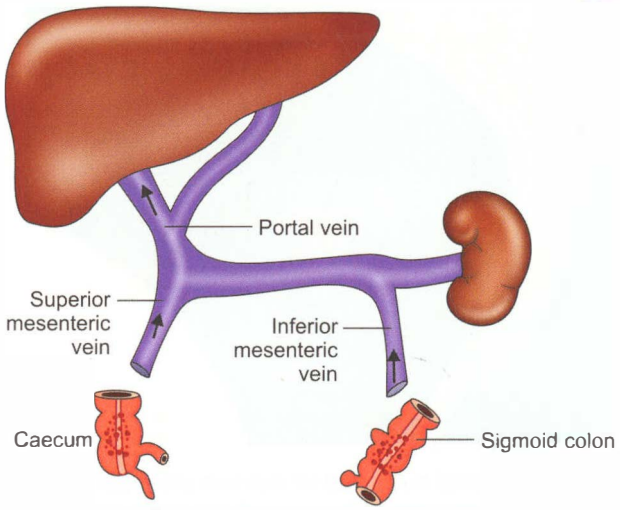


Fig. 24.6: Aetiopathogenesis of amoebic liver abscess

- The right branch of the portal vein is in direct line with the portal vein. Hence, by ***streamline phenomenon*** organisms reach the right lobe more often than the left lobe. The right lobe is also much bigger than the left lobe.
- In the right lobe, it is the posterosuperior surface which gets involved because it is extraperitoneal ***(bare area of liver)***. It has no peritoneal covering.
- After reaching the liver, the organism causes destruction of hepatocytes by releasing powerful ***cytolytic enzymes*** resulting in ***liquefaction necrosis***. It also causes aseptic thrombosis of blood vessels resulting in necrosis of liver tissue.
- At the same time, some RBCs[1] are also broken down. This causes ***anchovy sauce pus***, which is chocolate brown in colour, and is a mixture of broken down RBCs, hepatocytes, etc.
- **Green pus** is referred to pus mixed with bile, which is seen in a few patients.
- In majority of the cases, ***pus is sterile***. Secondary infection occurs in about 20 to 30% of the cases.
- Amoebae are rarely present in the pus but are present in the wall of the abscess cavity. The wall contains monocytes, plasma cells, lymphocytes and fibroblasts. Abscesses are multiple which fuse to form a single large abscess cavity in about 70% of the cases. Due to ***perihepatitis***, abscess gets fixed to the diaphragm resulting in immobility of the diaphragm. Liver abscess in the left lobe gets adhered to anterior abdominal wall.
- It is interesting to note that amoebic infection of gall bladder and bile does not occur because of ***deleterious effect of bile on amoebae***.

Clinical features (Fig. 24.8A)

- **Male alcoholics** are commonly affected, in the age group of 20–40 years. It is eight times more common in men.

[1]Absence of WBC is an important feature. Hence, regeneration of abscess cavity is complete without scarring.

Fig. 24.7: Intercostal bulge and tenderness are important features of amoebic liver abscess of the liver—these signs are not seen in acute cholecystitis

- Seen in patients with low socioeconomic status.
- Severe pain in the right hypochondrium is due to the enlarged liver. This stage is called stage of **Amoebic Hepatitis**. If USG is done, it may not demonstrate any abscesses but there may be many microabscesses. At this stage, there is low grade fever, weakness, anorexia, etc.
- High grade fever with chills and rigors develop if the stage proceeds to pyogenic liver abscess due to secondary bacterial infection of amoebic abscess.
- Thoracic symptoms such as nonproductive cough, pleurisy and right shoulder pain are common.

Signs

- Anaemia, emaciation, toxic look and an earthy complexion is present.
- Jaundice may be present if abscesses are multiple, due to compression of biliary radicles. However, it is rare (15%). It is of cholestatic variety.
- Liver is enlarged in the right hypochondrium, tender and soft (liver enlarges in upward direction) (Key Box 24.4).

KEY BOX 24.4

VERY TENDER LIVER

- Amoebic liver abscess
- Hepatoma
- Congestive cardiac failure

- **Intercostal tenderness** differentiates it from acute cholecystitis. Intercostal oedema can also be present (Figs 24.7 and 24.8A).

Investigations

1. **Total WBC count** may be increased if there is secondary infection.
2. **Stool examination** for ova and cysts of *Entamoeba histolytica* may be positive in 25% of cases.
3. **Serologic testing:** The indirect haemagglutination test is positive in 90–95% of patients with an amoebic abscess.
4. **Screening chest:** When the patient is asked to take a deep breath, right side of the diaphragm does not move due to inflammatory (**perihepatitis**) adhesions between liver and diaphragm. This is called **homolateral immobility of the diaphragm**. A small pleural effusion may also be present.
5. **Sigmoidoscopy** may demonstrate large, deep amoebic ulcers—**flask shaped**.
6. **Abdominal USG:** It is the investigation of choice.
   - To locate site of abscess and then to confirm diagnosis.
   - Ultrasound guided needle aspiration can also be done and biopsy of abscess wall should be taken.
   - Multiple abscesses can be made out.
7. **CT scan** can demonstrate an abscess cavity as a low density zone surrounded by peripheral hypodense zone due to inflammatory reaction.

Treatment

It can be classified into:

I. Conservative

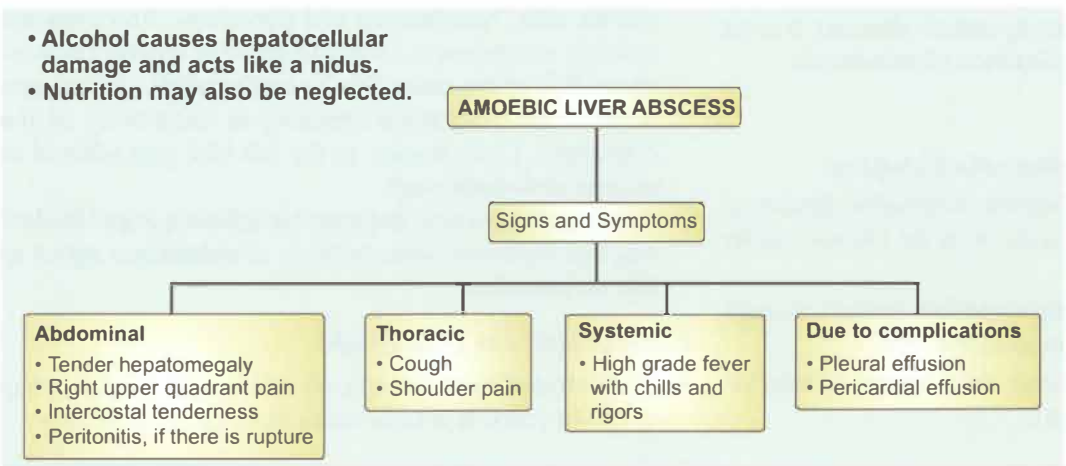


Fig. 24.8A: Clinical features

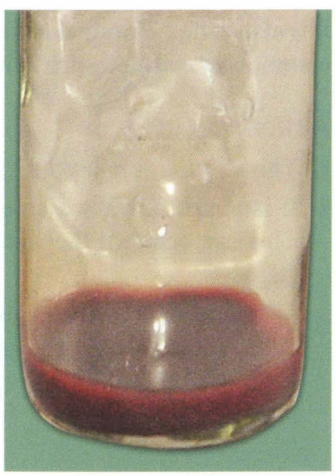


Fig. 24.8B: Anchovy sauce pus

II. Ultrasound-guided aspiration and pigtail drainage
III. Surgery—drainage

I. Conservative line of management

- It is indicated in ***amoebic hepatitis***. Tab. ***Metronidazole*** 400–800 mg, 3 times a day is given for 14 days.

  The only recognisable side-effect is metallic taste.
- If the condition does not improve, injection ***Emetine*** 1 mg/kg body weight to a total of 60 mg/day deep IM for a maximum of 6 days is given.

Side-effects and precautions during emetine therapy

- Systolic BP should be at least 100 mmHg.
- ECG should be recorded before, during and after the therapy.
- Cardiotoxicity in the form of arrhythmias can occur.
- Absolute bed rest during treatment (because of these complications, it is not used nowadays).
- Adequate hydration, rest, analgesics to relieve the pain.

Improvement can be seen within one to two days in the form of disappearance of pain, fever and return of appetite.

II. US-guided needle aspiration/pigtail catheter drainage (Key Box 24.5)

- It is indicated in cases of ***amoebic liver abscess***.
- Before it is aspirated, bleeding profile (BT, CT, PT) should be normal and injection vitamin K 10 mg, IM should be given for at least 3 days.
- US-guided aspiration is also the treatment of choice where ***metronidazole is contraindicated***, e.g. 1st trimester of pregnancy.
- It can be easily done under local anaesthesia
- Can be repeated, if pus recollects.

Typically it is anchovy sauce pus. Aspiration is followed by insertion of pig tail catheter.

Before removal of the catheter do a repeat ultrasound to check for residual pus.

KEY BOX 24.5

PIGTAIL CATHETER

- They are the catheter used to drain clear fluids from cavities such as pleural fluid, ascites, bile, etc.
- Different sizes are available 8 to 12 French
- Radio-opaque polyurethane catheter
- Length – 20 or 30 cm
- Large oval holes in pigtail for maximum drainage
- Thus, maximum efficiency is when 'transudate' pleural effusion is drained, than empyema, haemothorax or anchovy sauce pus.

The clinical response to the aspiration may be observed as subsiding of fever (Figs 24.8B to 24.11).

Complications

1. Bleeding—rare
2. Incomplete aspiration results in leakage of pus and bile into peritoneal cavity which may produce peritonitis. Hence, prophylactic antibiotics need to be given before and after the procedure along with metronidazole therapy.

III. Surgery (open drainage) and laparoscopic

Indications

1. Failure of US-guided needle aspiration.
2. Ruptured amoebic liver abscess with amoebic peritonitis.

Procedure

- Laparotomy is done first. Abscess cavity is identified. Contents are evacuated, a thorough peritoneal wash is given and a self-retaining **Malecot's catheter (any tube drain)** is introduced into the abscess cavity, brought outside and connected to a bag.
- With the advent of metronidazole, **Amoebiasis Cutis is rarely seen**. Hence, catheter can be safely placed inside the cavity and brought out. Malecot's catheter is now being replaced by other tube drains.
- Postoperatively for 3–5 days, necrotic liver tissue, anchovy sauce pus and blood drain outside.

Fig. 24.9: Pigtail catheter drainage of amoebic liver abscess—it is a nonoperative method of treatment

Fig. 24.10: Ultrasound is done to check residual abscess cavity

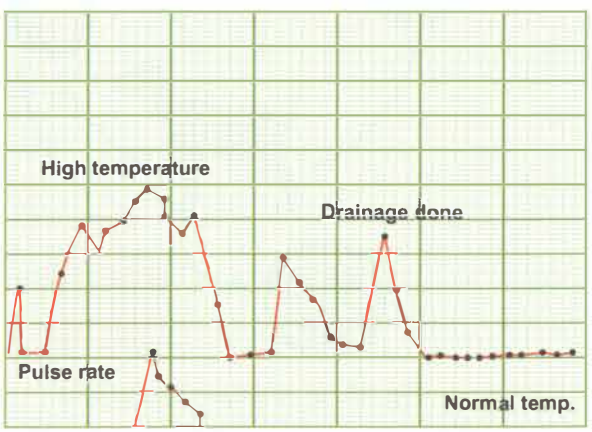


Fig. 24.11: Temperature chart—once the drainage is done, temperature comes back to normal

- Once the drainage becomes minimal, Malecot's catheter is pulled out.
- Same procedure can be done by laparoscopic method.

Complications of amoebic liver abscess

- **Amoebic peritonitis**, resulting in acute abdomen with shock. It has to be treated like any peritonitis—laparotomy, drainage of pus and drain the abscess cavity to outside (possibility of ***amoebiasis*** cutis is still present but rare).
- Rupture into pleural space causing ***pleural effusion***.
- Rupture into the bronchus resulting in coughing out anchovy sauce (may be a natural cure)—***bronchopleural fistula***.
- **Amoebic pericardial effusion** occurs due to rupture of left liver lobe abscess into pericardial space.

HYDATID CYST OF THE LIVER

The disease is caused by *Echinococcus granulosus*, transmitted by dogs which are the chief mediators (host) and man is the intermediate host (Fig. 24.12). After swallowing the ova, they penetrate gastric mucosa[1], reach retroperitoneal structures, penetrate portal vein directly and then enter into liver. Having reached liver, the organisms grow and develop their own protective layer and form hydatid cyst.

Layers of hydatid cyst (Fig. 24.13)

1. **The adventitia (pseudocyst)[2]:** This is the fibrous layer derived from the liver tissue. It is the reaction of liver to the parasite. It is adherent to the liver and cannot be separated—pericyst.

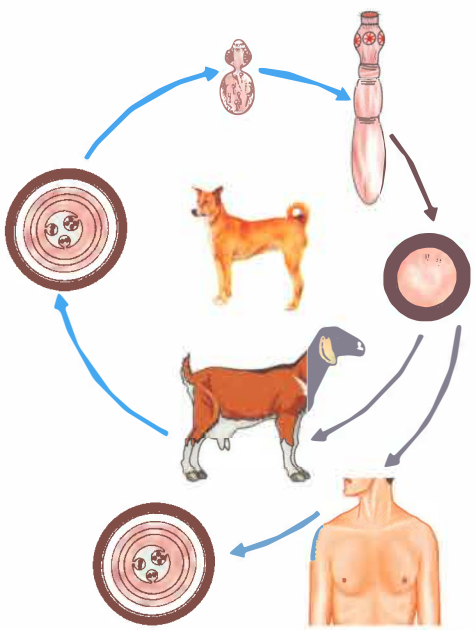


Fig. 24.12: Life cycle hydatid cyst

[1]Reason why Hydatid ova do not produce lesions in the intestine.
[2]It is the protective coat given by the host to the ghost.

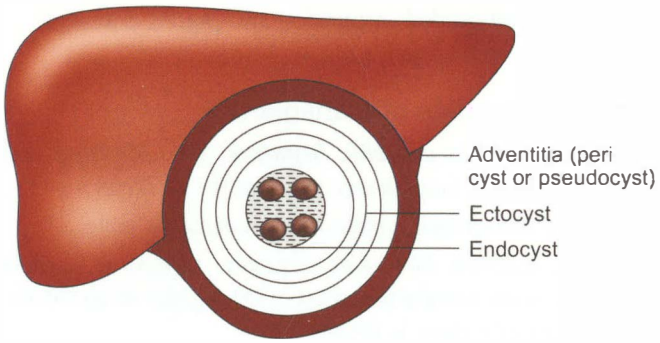


Fig. 24.13: Layers of hydatid cyst

2. **The ectocyst (laminated membrane):** It is white and elastic and is produced by endocyst. It is this layer which gets peeled off at surgery.
3. **The endocyst:** This is called germinal epithelium and it is the innermost part of hydatid cyst. It secretes hydatid fluid inside and ectocyst outside. Within the hydatid fluid, the 'brood capsules' develop within which the scolices of *Echinococcus granulosus* develop.

Clinical features

- It can be silent—without any symptoms throughout life, accidentally discovered on routine examination.
- Dragging pain in the upper abdomen due to hepatomegaly.
- Liver is enlarged, has a smooth surface, round borders and is nontender.
- **Typical hydatid thrill** can be present on rare occasions. Hydatid thrill is demonstrated by 3-finger method. Keep 3 fingers over the liver, percuss over the middle finger and get the impulse by other 2 fingers (fluid thrill).
- Patient may present as an emergency with severe abdominal pain following minor trauma.
- May present as an emergency with features of anaphylatic shock without any obvious cause.

Investigations

1. **USG** can detect the cyst, localise the cyst and is used for aspiration purposes (details later—PAIR).
   WHO—Informal working group of echinococcosis (WHO—IWGE) (USG classification)
   - **Group-1:** Active group—cyst larger than 2 cm and often fertile
   - **Group-2:** Transition group—cyst starting to degenerate because of host resistance or treatment; may contain viable protoscolices
   - **Group-3:** Inactive group—degenerated partially or totally calcified cyst; unlikely to contain viable protoscolices
2. **Plain X-ray abdomen** may demonstrate **speckled** calcification.

3. **CT scan** may be necessary in selected cases (Figs 24.14 and 24.15). The cyst which is superficial and has reached surface, should be operated upon.
4. **ERCP** if there is obstructive jaundice—in such cases, a wide sphincterotomy should be given so as to allow free drainage of the hydatid contents into the duodenum.
5. **Casoni's intradermal test:** Sensitivity and specificity of this test is low and hence, **no longer used**.
6. ELISA and immunoelectrophoresis may point towards the diagnosis.

Treatment

It can be discussed under following headings:

I. Conservative

1. Calcified cysts are dead cysts. They are left alone.
2. Symptomless, small hydatid cyst can be left alone. Once symptomatic, or if the size is more than 5 cm, they may be treated.

II. Medical treatment (Key Box 24.6)

- Albendazole 8 mg/kg or 400 mg BD is given for 21 days followed by drug holiday for 2 weeks.

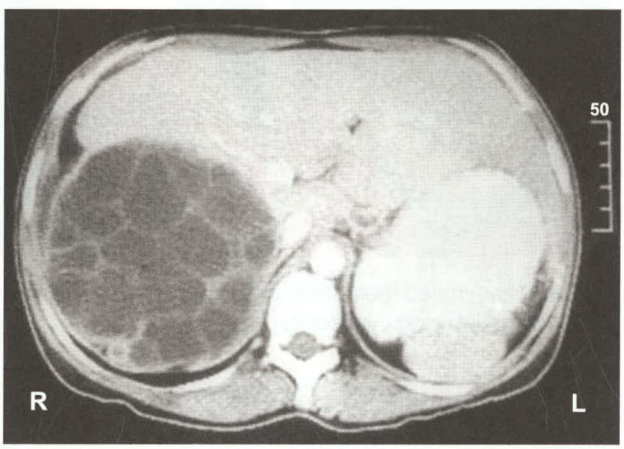


Fig. 24.14: CT scan of the liver showing classical cart wheel pattern—coronal section

Fig. 24.15: CT scan of the liver showing classical cart wheel pattern—sagittal section

If no improvement occurs maximum of 3 such cycles can be given.

- Watch for neutropenia

III. Surgery

There are different types of surgeries for hydatid disease of the liver which have been summarised below. Indications are given in Key Box 24.7.

KEY BOX 24.6

INDICATIONS FOR MEDICAL TREATMENT

- Disseminated hydatid cyst
- Inaccessible for surgery (deep seated, multiple, recurrent)
- Patient unfit for surgery
- Contamination of peritoneal cavity at surgery

KEY BOX 24.7

INDICATIONS FOR SURGERY

- Symptomatic cyst
- Asymptomatic patient with cyst > 5 cm, noncalcified
- Infected cyst (Figs 24.20 and 24.21)

Principles of surgery

1. **Laparotomy** and **isolation of the cyst** from peritoneal cavity by packs dipped in hypochlorite solution.
2. Aspirate the contents and inject **scolicidal agents** such as savlon or hypertonic saline.
3. Incise the cyst, **peel off the laminated membrane** by using sponge-holding forceps (Figs 24.16 to 24.21).
4. An attempt to remove adventitial layer may result in bleeding. It need not be removed.
5. At surgery, all laminated membrane, endocyst has to be removed. Perfect haemostasis must be achieved. Any bile leak to be sutured. Postoperative significant bile leak is managed by ERCP stenting.

Precautions

1. Albendazole should be given before surgery.
2. Avoid spillage into the peritoneal cavity to avoid peritoneal hydatid.
3. **Injection hydrocortisone** 100 mg IV before, during and after surgery to avoid anaphylactic shock.

Different types of surgical procedures

They have been summarised in Key Box 24.9. However, excision of the cyst leaving the adventitial layer is the most commonly performed surgery (Figs 24.19 to 24.21).

IV. Percutaneous drainage

- It is done after taking all necessary precautions/equipment with all emergency drugs and with CT/US guidance.

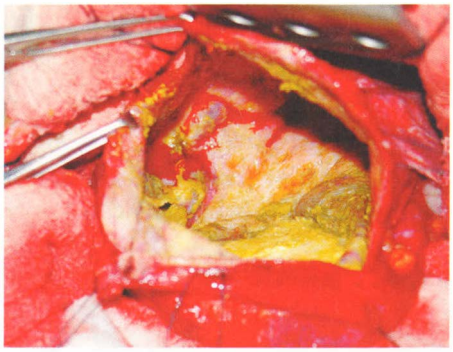


Fig. 24.16: Infected ruptured hydatid cyst— observe green colour due to biliary communication

Fig. 24.17: Pearl-like daughter cysts (very precious!!)—Hydatid means dew drop in Latin and watery vesicle in Greek

Fig. 24.18: Daughter cysts—it is important to clear all these to prevent recurrence

(*Courtesy:* Dr Geetha R, Dr Ankur Sharma, Dr Rajesh Nair, Department of Surgery, KMC, Manipal)

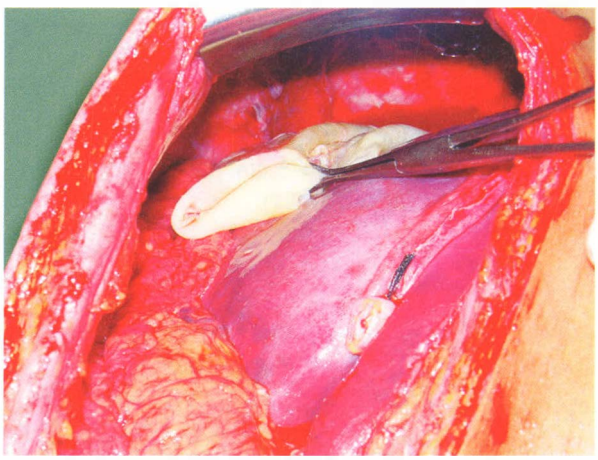
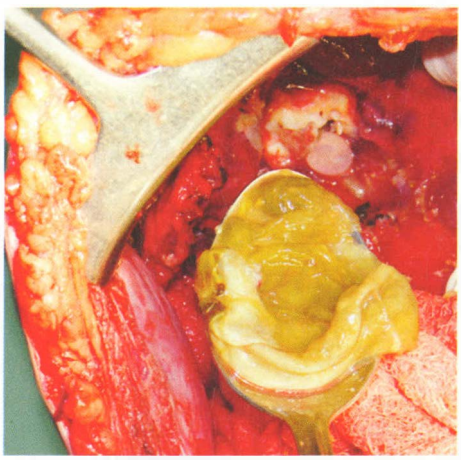


Fig. 24.19: Laminated membrane removed at surgery with sponge-holding forceps



Figs 24.20 and 24.21: Laminated membrane and daughter cysts are being removed. You can see the infected cyst cavity

- It is also called **PAIR**—**P**uncture, **A**spiration, **I**njection and **R**easpiration.

Indications (Fig. 24.22)

- Unilocular uncomplicated cyst
- Poor surgical candidate
- Previous multiple abdominal surgeries
- Relapse following surgery

Contraindications to PAIR (Fig. 24.23)

- Multiple septal divisions
- Communicating cysts
- Dead or inactive cysts
- Laparoscopic approach for these procedures are being done.

Complications of hydatid cyst

1. **Rupture** of the cyst into the peritoneal cavity can occur due to trauma (rare) and may result in anaphylactic shock. It should be treated accordingly with injection hydrocortisone and laparotomy.

KEY BOX 24.8

SCOLICIDAL AGENTS	SIDE-EFFECTS
• Hypertonic saline	• Hypernatraemia
• Chlorhexidine	• Acidosis
• Alcohol (80%)	• Cholangitis
• Sodium hypochlorite	• Hypernatraemia

KEY BOX 24.9

SURGICAL PROCEDURES

- **Excision:** It is the most commonly performed operation.
- **Cystopericystectomy:** Here, the cyst is excised in a plane outside adventitial layer. It results in excessive bleeding but it removes the cyst completely.
- **Capitonnage:** If omentum is not available for closing the cavity, redundant cyst wall can be infolded into the cyst cavity and sutured.
- **Hepatic resection** in good centres when there are proper indications.

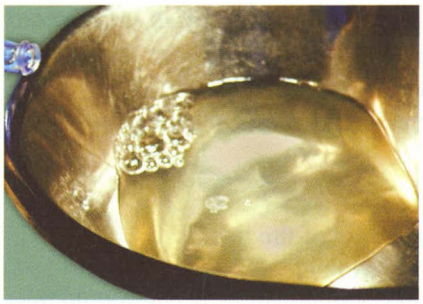


Fig. 24.22: Clear fluid being aspirated as a part of PAIR treatment

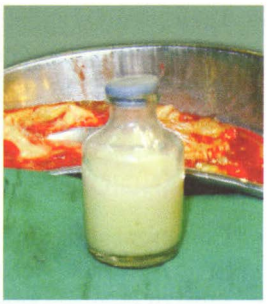


Fig. 24.23: As the aspirated fluid was turbid, open drainage was done in this patient

- Rupture can result in implantation of active cysts within peritoneal cavity and they grow within. Rupture through diaphragm can cause **empyema**. It can also rupture into biliary radicles and can cause obstructive jaundice.

2. **Jaundice** due to cysts within biliary tree or due to a large cyst compressing biliary ducts. ERCP followed by endoscopic removal of hydatid cysts from the common bile duct followed by sphincterotomy should be done.
3. **Suppuration** is rare due to the tough tunica adventitia.
4. **Calcification** of the cyst wall in long-standing cases (Fig. 24.24).

Other type of organism is ***Echinococcus multilocularis.*** This parasitic infestation is uncommon in our country but it is more severe (Key Box 24.10).

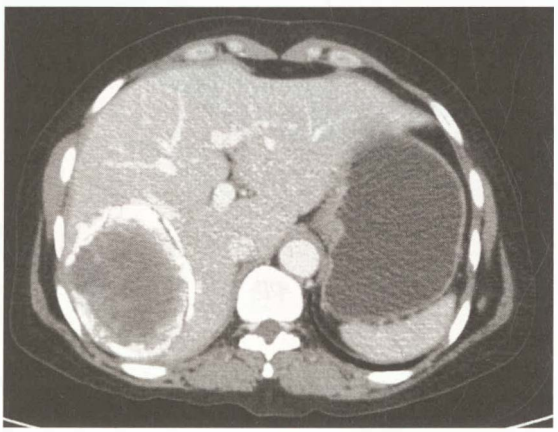


Fig. 24.24: CT of the liver—hydatid calcification

KEY BOX 24.10

ECHINOCOCCUS MULTILOCULARIS

- Alveolar echinococcosis
- It can cause **malignant hydatid disease** (misnomer) Difficult to treat. Hence, the name
- Needs resection
- It causes poorly demarcated honeycombed cystic specimen
- It infiltrates liver and invades vascular system

OTHER CYSTIC DISEASES

SIMPLE CYSTIC DISEASE

- When the cyst is thin walled, unilocular and regular it is usually a simple cyst (Figs 24.25 to 24.27).
- The host response is usually not present (unlike hydatid cyst).
- Incidentally discovered by ultrasound examination.
- Clinically, if they are big, liver will be palpable with smooth surface and round borders. It is nontender.

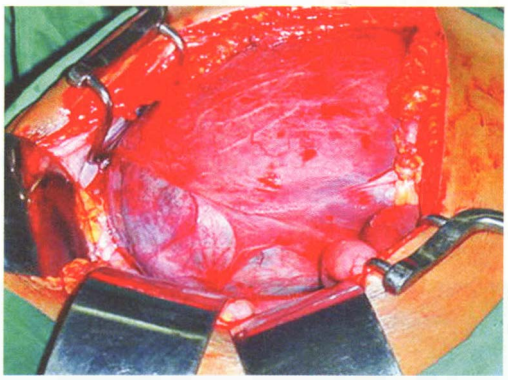


Fig. 24.25: Simple cyst at surgery. Observe the thin wall with slight bluish colour

Fig. 24.26: De-roofing has been done

Fig. 24.27: Colour of the fluid—slightly greenish

- Asymptomatic cysts do not require any treatment.
- Ultrasound guided aspiration, laparoscopic de-roofing and open surgical de-roofing are the methods of treating the cyst.
- Differential diagnosis include hydatid cyst, polycystic disease.

POLYCYSTIC LIVER DISEASE

- Congenital abnormality
- Cysts occur within liver, pancreas and kidney (principally)
- Cysts are asymptomatic
- Cysts do not produce liver cell failure (unlike polycystic kidneys).
- Cyst haemorrhage may be the presenting feature with right hypochondriac pain mimicking cholecystitis.
- CT scan and ultrasound are investigations.
- Cyst aspiration (PAIR like hydatid cyst), decompression may rarely be required in large painful lesions (laparoscopic). ***Observe 7 Cs of polycystic liver*** (Fig. 24.28).

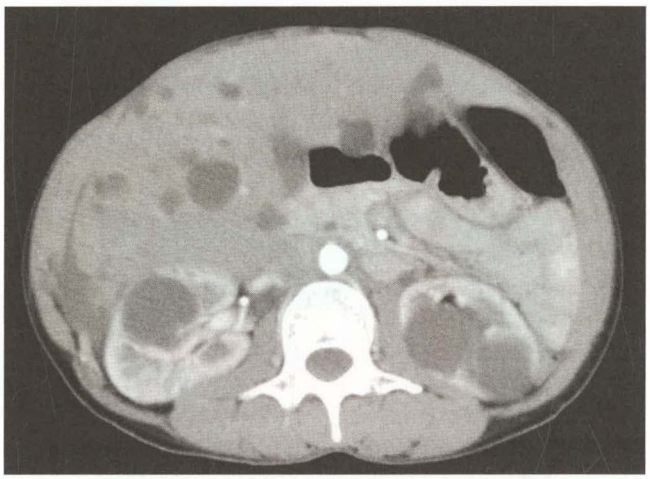


Fig. 24.28: CT scan showing polycystic liver and polycystic kidney

BENIGN TUMOURS OF THE LIVER

- These are not uncommon tumours.
- They are more frequently diagnosed now with frequent use of ultrasound. Majority of them are asymptomatic and do not require specific treatment.
- Their removal should be attempted only by an experienced surgeon. Hepatic adenoma and focal nodular hyperplasia are compared in Table 24.2.
- They have a rare potential for malignant change.

HAEMANGIOMA

- **Commonest benign tumour** of the liver
- Majority (75%) occur in **women**
- Most of them are **solitary, subcapsular**, occur in right lobe of liver
- It may be associated with **cavernous haemangioma** at some other sites such as head and neck region. Clinically, it is difficult to diagnose as it presents as hepatomegaly.
- **Kasabach-Merritt syndrome: Thrombocytopaenia and consumptive coagulopathy.**
- Sometimes it may cause cardiovascular failure due to sequestration of blood.
- Ultrasound/CT scan can diagnose their location, number (single or multiple) or presence of any other complications associated with that (thrombosis, infection). MRI is a better investigation.
- Haemangioma **bigger than 8 cm** have **chances of rupture.**

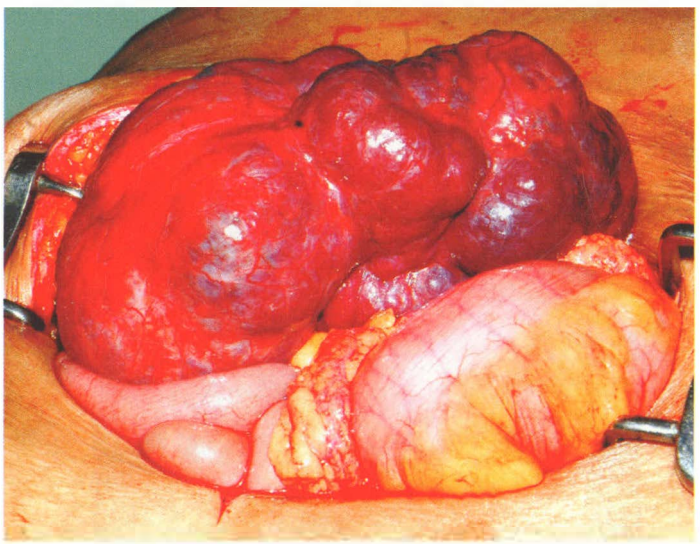
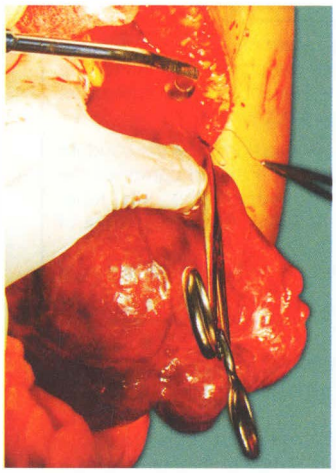


Fig. 24.29: Haemangioma arising from the inferior surface of the liver, is exposed at surgery

Table 24.2 Comparison of hepatic adenoma and focal nodular hyperplasia

Hepatic adenomas	Focal nodular hyperplasia
• Occur in young women	• Occurs in young women
• Occur due to oral contraceptive pills (OCPs)	• Occurs due to oral contraceptive pills
• Usually **solitary**	• Can be **multiple**
• Haemorrhage, rupture, necrosis can occur	• Haemorrhage, rupture, necrosis do not occur
• Can undergo malignant change	• **Rarely, malignant** change can occur
• If it is due to OCPs, it can regress after discontinuing OCPs. Otherwise, adenomas need to be resected	• Generally does not regress
• Kupffer cells are **not present** in hepatic adenomas	• In addition to hepatocytes, it also contains Kupffer cells



Figs 24.30 and 24.31: As it had a very narrow strip of the liver tissue, it was easily excised

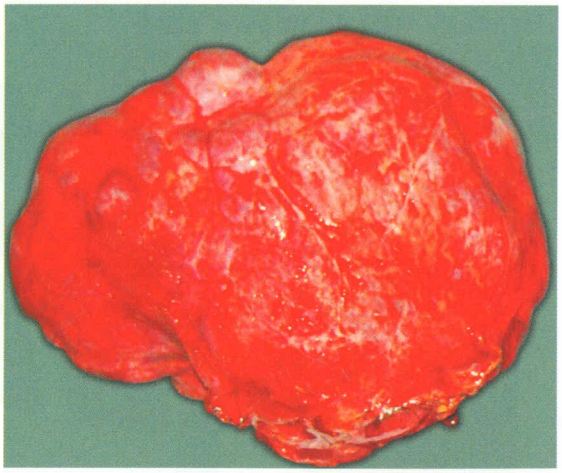


Fig. 24.32: Resected specimen

Treatment

1. **Trans-arterial embolisation (TAE):**
- Indicated in large haemangiomas which are unresectable.
- Large haemangiomas on the inferior surface of the liver are ideally suitable for the treatment (Figs 24.29 to 24.32).
- Embolic materials
  (A) Temporary: Gel foam
  (B) Permanent: Steel coils, polyvinyl alcohol, isobutyl cyanoacrylate.
- Polyvinyl alcohol particles 300–500 μm in size are used often.

2. They can be resected or can be enucleated.

Caution: Before putting a needle into the space occupying lesion in the liver, make sure it is **NOT a haemangioma**.

Differential diagnosis

- Hepatoma, liver cysts, liver abscess

Other benign tumours are hepatic adenomas and focal nodular hyperplasia.

PRIMARY LIVER CANCER—HEPATOMA OR HEPATOCELLULAR CARCINOMA (HCC)

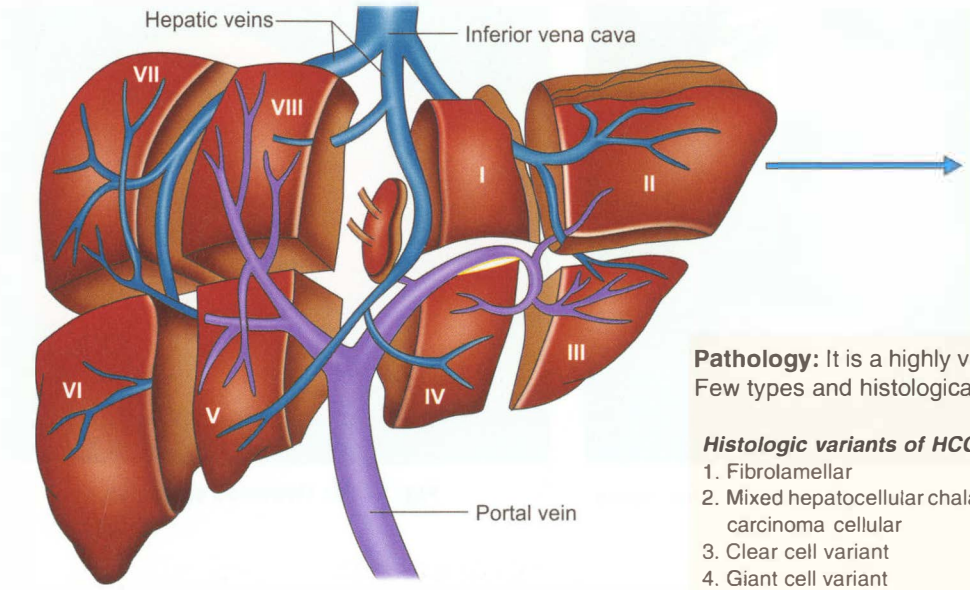
- Before discussion on HCC, one has to have a knowledge about segmental anatomy of the liver, which forms the basis of hepatic resection.
- Even though the details about hepatic resection may not be required for undergraduate students, it is desirable to know about the segmental anatomy to have a better understanding about hepatic resection.

SEGMENTAL ANATOMY OF THE LIVER

- The liver is divided into two lobes by the main portal fissure, which is also called **Cantlie's line**. This line extends from the gall bladder fossa to the left side of IVC and is inclined at an angle of 75° to the horizontal plane. The main portal fissure is a constant feature. Each lobe is equal in size.
- The **right portal fissure** divides the right lobe into an anteromedial and posterolateral sector. The right hepatic vein courses along this fissure. This fissure is inclined at an angle of 40° with the horizontal.
- The left portal fissure divides the left lobe into an anterior and posterior sector, and it is in this fissure that the left hepatic vein courses.
- The liver is further divided into segments, which represents the smallest anatomic unit of the organ. In the right lobe, each of the two sectors is divided into two segments: The anteromedial sector and posterolateral sector. The anteromedial sector is divided into segment V anteriorly and segment VIII posteriorly, while the posterolateral sector is divided into segment VI anteriorly and segment VII posteriorly.
- In the left lobe, the anterior sector is divided into segment IV (or quadrate lobe) and the segment III. The posterior sector is comprised of only one segment, segment II. The Spigelian lobe or segment I is considered as an ***autonomous*** segment. It is also labelled ***caudate lobe***.
- Each segment has its own identifiable portal vein, hepatic arterial supply, bile duct and hepatic veins. All these segments can be removed individually. However, segment I receives tributaries from both the right and left branches of portal vein and hepatic artery. ***However, hepatic venous drainage is independent and may terminate directly into IVC*** (Fig. 24.33).

Introduction

- Hepatocellular carcinoma is uncommon in United States but more in Asians. It arises from hepatocytes.
- When it arises from biliary epithelial cells,it is intrahepatic cholangiocarcinoma.
- In neonates, it is hepatoblastoma as it resembles foetal liver.



Segmental anatomy of the liver with the 8 segments, based on portal vein blood supply and hepatic venous outflow as designated by Couinaud.

Segment I is also known as the caudate lobe, segments II and III comprise the lateral segment of the left lobe, segment IV is the medial segment of the left lobe.
Segments V to VIII comprise the right lobe.

Fig. 24.33: Segmental anatomy of the liver

Pathology: It is a highly vascular tumour and fatal if left untreated. Few types and histological features are given below

Histologic variants of HCC
1. Fibrolamellar
2. Mixed hepatocellular chalangios-carcinoma cellular
3. Clear cell variant
4. Giant cell variant
5. Childhood HCC
6. Carcinosarcoma

*Refer Table 24.3 in the next page

Pathology: Histology types
- It is a highly vascular tumour with indistinct margin.
- **Hanging type:** It is attached to the liver by a small stalk—easily resected.
- **Pushing type:** A tumour pushing the blood vessels. It is also resectable.
- **Infiltrative type:** Difficult to resect because of infiltration into surrounding structures.
- Hence, in cirrhotic patients, resection is not advisable.

Aetiology of HCC

1. **Hepatitis B virus (HBV):** High titres of hepatitis B surface antigen (HBsAg) was found in 50 to 60% of patients with HCC. Its incidence is higher in chronic carriers and it is increased if patients consume alcohol, aflatoxin and tobacco.
2. **Hepatitis C virus (HCV):** HCV infection induces cirrhosis and increases the incidence of development of HCC.
3. **Cirrhosis:** It is a definite premalignant condition. A palpable hard liver in a cirrhotic patient suggests development of HCC.
4. **Aflatoxin consumption** (a fungus found in infected butter yellow, bread) is responsible for increased incidence of HCC as in Mozambique.
5. It is 4 times more common in Asians and two times in Hispanics. Less common in Caucasians.
6. **Oral contraceptive pills** are known to cause adenoma and there is a rare chance of adenoma turning into HCC.
7. **Other risk factors:** Heavy alcohol consumption, cigarette smoking, diabetes mellitus, haemochromatosis are the other factors responsible for HCC. ***Wilson's disease is rarely associated with increased risk of HCC.***
8. Hereditary tyrosinemia is a metabolic disorder characterised by deficiency of enzyme fumaryl acetoacetate resulting in, accumulation of toxic tyrosine metabolites—can progress to HCC.
9. **Other causes:** Type I Glycogen storage disease and familial polyposis coli Alpha-1 Antitrypsin deficiency, Budd-Chiari syndrome

Clinical features

- **Age group:** Highest incidence is found after 50 years of age. It is rarely seen in children also.

PEARLS OF WISDOM

Thus almost 80% of the cases are due to underlying chronic hepatitis B and C virus infection.

- Sex: ***Male alcoholics*** are commonly affected
- A ***slow growing*** mass in the right hypochondrium. It is the enlarged liver with an irregular surface and is hard in consistency. Evidence of cirrhosis may be present.
- It can be a ***rapidly growing*** neoplasm, highly vascular and clinically palpable as a soft to firm pulsatile mass with or without a bruit.
- Rapid development of ***anorexia, asthaenia, anaemia and loss of weight.***
- Jaundice is not common unless the tumour arises in a cirrhotic liver in hepatocellular failure.
- Worsening of portal hypertension.
- **Low grade pyrexia** is common. It is due to tumour necrosis.
- Liver being an important organ of metabolism, **hypoglycaemia** is found in 10% of the patients.
- Ascites
- Triad of abdominal pain, weight loss and abdominal mass is the most common clinical presentation.

Spread of hepatoma

1. **Direct infiltration** of the neighbouring structures such as diaphragm results in hiccoughs due to irritation of phrenic nerve.

Table 24.3 Comparison of HCC and fibrolamellar carcinoma

	Hepatocellular carcinoma	Fibrolamellar carcinoma
1. Age (years)	50–60	20–30
2. Sex	More common in males	Equal in both sexes
3. Aetiology	Cirrhosis	Does not arise in a cirrhotic liver
4. Involvement of lobe	Right lobe	Left lobe
5. Gross pathology	Solitary or nodular, central necrosis is common	Solitary or nodular, central necrosis is not common
6. Microscopy	Basic structure is trabecular pattern with acidophilic cytoplasm and large nuclei	Deeply eosinophilic hepatocytes, fibrous stroma is arranged in the form of thin hyalinised bands in layers. Hence the name, **fibrolamellar**
7. Resectability rate	Less	More
8. Prognosis	Not good	Good

2. **Lymphatic spread:** Lymph nodes at the hilum (porta hepatis) are involved first. Later, mediastinal nodes and left supraclavicular nodes (Virchow's node) are involved.
3. **Blood spread** causes massive malignant pleural effusion by dislodgement of tumour emboli into inferior vena cava because of spread through the hepatic veins. Vertebral involvement follows soon.
4. **Haemoperitoneum** due to spread involving peritoneal surface or due to rupture of an enlarging hepatoma may occur.

Investigations

1. **CBP:** Hb% is usually low.
2. **Liver function tests** may show hepatocellular failure in the form of high bilirubin, low albumin and high globulin levels.
3. **Chest X-ray:** CT chest pulmonary metastasis
4. **Abdominal USG:** Findings
   - Diffuse distortion of hepatic parenchyma and a well-circumscribed hyperechogenic mass suggests HCC.
   - **Mosaic pattern** of the tumour with thin halo and lateral shadows, **nodule in nodule pattern** with separating fibrous septa and **posterior echo enhancement**.
   - **Tumour thrombi** in portal vein, hepatic vein or IVC.
5. **Contrast enhanced CT of the abdomen**[1]**:** It appears as a hypodense mass with internal mosaic architecture in the early phase and hypodense lesion with enhanced fibrous capsule in late phase. Guided FNAC can be done to confirm the diagnosis. A small fear of tumour embolisation *via* portal venous radicles exists and is a complication. Enhancement in arterial phase and washout in delayed portovenous phase is diagnostic of HCC (Figs 24.34 and 24.35).

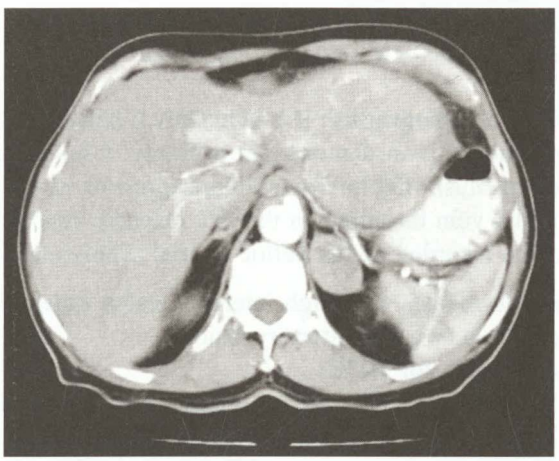


Fig. 24.34: CT scan showing a large hypodense mass lesion in the left lobe

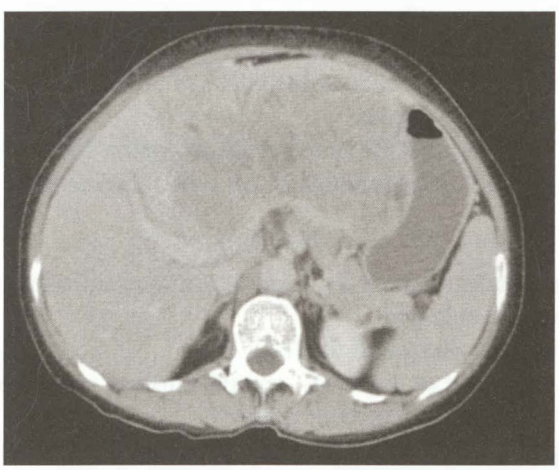


Fig. 24.35: CT scan showing a large hepatoma displacing the portal vein branches

[1]Triple phase—Precontrast, early vascular or arterial, portal phase and delayed phase—multislice spiral CT is the gold standard investigation for liver tumours. If clinical, radiological and biochemical features strongly suggest HCC, there is no need to prove histopathological diagnosis by FNAC.

6. Alpha-fetoprotein

- It is a foetal antigen which disappears after birth. Hence, it is not measurable in normal persons.
- Values above 20 ng/ml are suggestive of hepatoma.
- Levels above 400 ng/ml in a cirrhotic patients with hypervascular mass larger than 2 cm in diameter is diagnostic.
- α–fetoprotein levels indicate size of the tumour, invasion and even recurrence.
- In patients who are not operated, levels reflect response to treatment (Key Box 24.11).

KEY BOX 24.11

INCREASED ALPHA-FETOPROTEIN LEVELS

1. **Hepatoma—Hepatoblastoma**
2. Carcinoma stomach
3. Carcinoma pancreas
4. Embryonal cell carcinoma of testis

Not increased in fibrolamellar carcinoma

7. Selective angiography: It is done only if hepatic resection is planned. It can demonstrate a highly vascular tumour (tumour blush due to neovascularisation) or involvement of portal vein thrombus in the IVC. Hence, venous phase must be recorded. It can define arterial supply also.

8. MRI: It appears as a high intensity lesion (adenomatous hyperplasia as a low intensity lesion).

9. Liver biopsy for HCC:

- It is done only in unresectable cases or metastatic disease before starting other treatment such as chemotherapy, etc.
- The images from contrast CT scan and MRI and raised levels of α-fetoprotein—almost always suggest HCC. Hence, biopsy is not done.
- Also rises of haemoperitoneum and peritoneal implantation of tumour cells present.
- Core-biopsy has increased risks of all the complications mentioned above.
- With normal INR, fine needle aspiration can be done in selected cases (clinical trials) with minimal risk.

Differential diagnosis (Key Box 24.12)

- Secondaries in the liver—nodular surface and usually both lobes are enlarged.
- Polycystic disease of the liver
- Hydatid cyst of the liver—smooth surface, round borders.

Refer TNM staging of liver cancer

KEY BOX 24.12

DIFFERENTIAL DIAGNOSIS

- Fibrolamellar carcinoma
- Secondaries in the liver
- Haemangioma
- Focal nodular hyperplasia
- Polycystic liver

TNM STAGING LIVER CANCER

Tumour

T0 No primary tumour
T1 Solitary tumour without vascular invasion
T2 Solitary tumour with vascular invasion/multiple tumours equal to or less than 5 cm
T3 Multiple tumours more than 5 cm/tumour invading major branch of portal or hepatic veins
T4 Tumour with direct invasion of adjacent organs other than gall bladder and visceral peritoneum

Nodal status

N0 No regional lymph node metastasis
N1 Regional nodes are involved

Metastasis

M0 No distal spread
M1 Distant spread present

CHILD-TURCOTTE-PUGH (CTP) SCORE

Classification of hepatocellular function in cirrhosis

• Points	1 point each	2 points each	3 points each
• Bilirubin (μmol/L)	< 34	34–50	> 50
• Albumin (g/L)	> 35	25–35	< 25
• Ascites	None	Easily controlled	Poorly controlled
• Encephalopathy	None	Grade I–II	Grade III–IV
• INR	< 1.7	1.7–2.2	> 2.2

A—5–6 points B—7–9 points C—10–15 points

INR—international normalised ratio

Treatment of hepatocellular carcinoma

1. Resection is the best possible treatment for hepatocellular carcinoma. Up to 3 segments of the liver can be resected. Rest of the liver will be enough to maintain life provided remaining liver is healthy/noncirrhotic (Tables 24.4 and 24.5).

- Functional liver remnant (FLR) should be > 20% of standardised total liver volume after resection.
- The remnant liver should have vascular inflow, venous outflow, biliary drainage.
- Two contiguous liver segments must be left behind (Figs 24.36 to 24.41).

2. Systemic chemotherapy using intravenous doxorubicin has been tried with some success in the postoperative period. Intra-arterial 5-FU has also been tried.

3. Transcatheter arterial embolisation (TAE or TACE) by introducing gel foam into the branches of hepatic artery produces some amount of tumour necrosis. If this arterial embolisation is combined with chemotherapeutic agents

Table 24.4 Various segments for liver resection

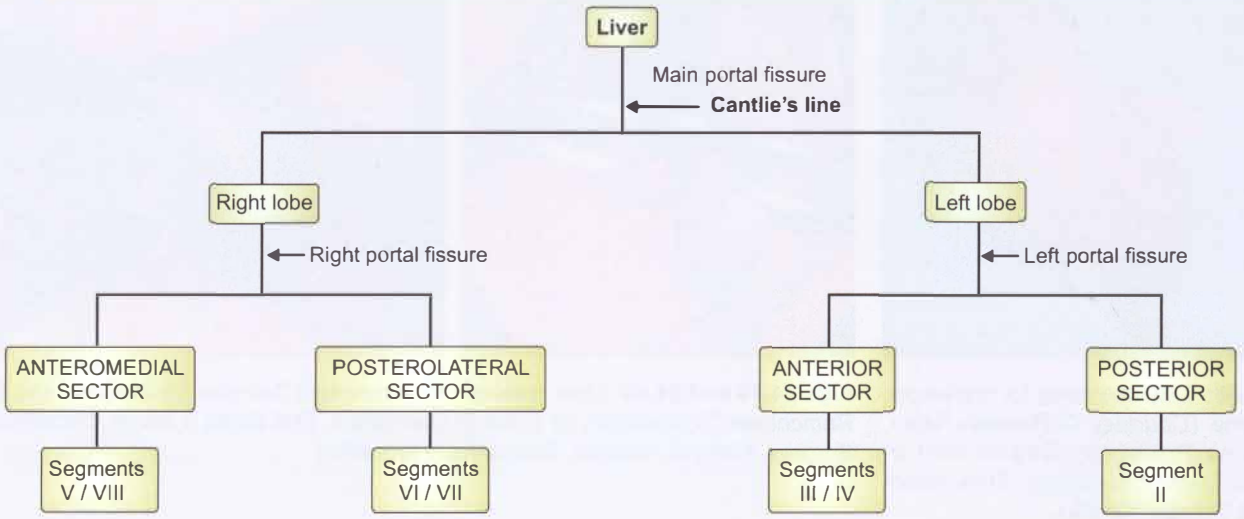
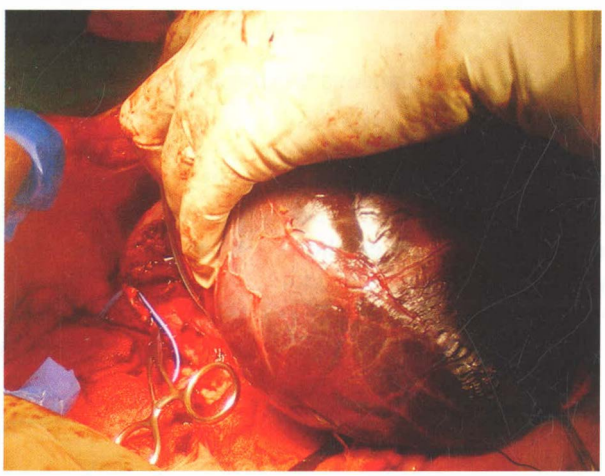


Table 24.5 Types of hepatic resections

Hepatic resection	Nomenclature
1. Resection along the main portal fissure, separating right and left lobes of the liver	• Right or left lobectomy
2. Resection of entire right lobe and the medial segment of left lobe	• Right extended lobectomy or trisegmentectomy
3. Removal of segments II and III	• Left lateral segmentectomy
4. Removal of a single segment	• Unisegmentectomy
5. Removal of 2 or more segments	• Plurisegmentectomy
6. Removal of segments VI and VII	• Right lateral sectorectomy
7. Removal of a small portion of liver, entirely within a single segment or transferring segmental plane	• Wedge resection



Figs 24.36 and 24.37: Liver resection for adenoma (*Courtesy:* Dr Prasad Babu TLVD, Dr Ramcharan Thyagarajan, Dr Srikanth Gadiyaram, Prof Sadiq S Sikora, Department of GI Surgery, Manipal Hospital, Bengaluru, Karnataka)

such as doxorubicin, results are better because the tumour has prolonged exposure to the drug. This is called **trans arterial chemoembolisation**. This method is followed only as a palliative procedure.

Contraindications

- Serum bilirubin > 3 mg/dl
- Tumour thrombus in main portal trunk
- Early HCC

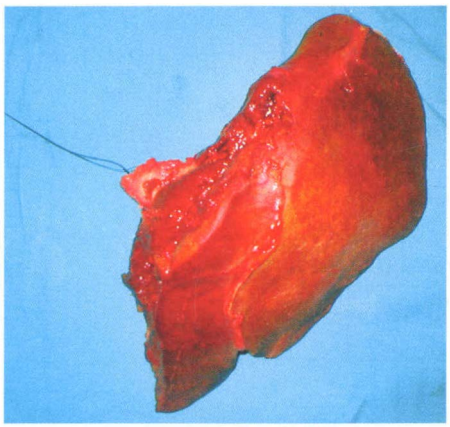
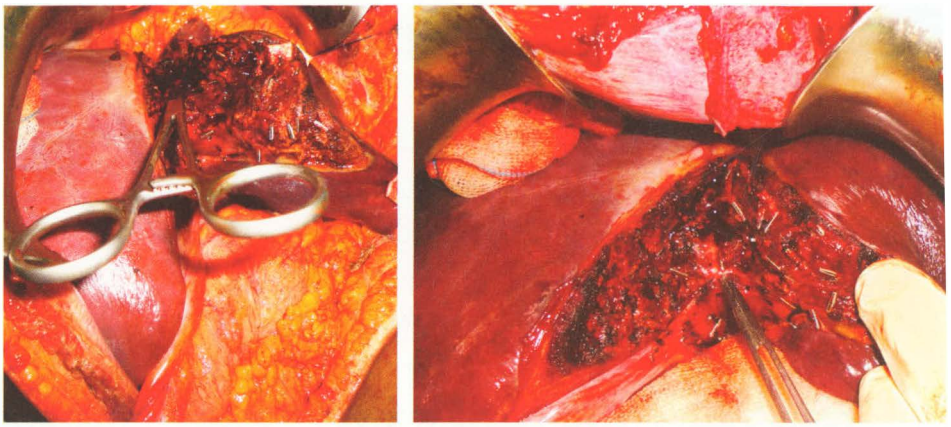


Fig. 24.38: Left lobectomy for cholangiocarcinoma (*Courtesy:* Dr Ramesh Rajan, Associate Professor, Department of Surgical Gastroenterology, Trivandrum Medical College, Kerala)



Figs 24.39 and 24.40: Liver resection for adenoma (*Courtesy:* Dr Prasad Babu TLVD, Dr Ramcharan Thyagarajan, Dr Srikanth Gadiyaram, Prof Sadiq S Sikora, Department of GI Surgery, Manipal Hospital, Bengaluru, Karnataka)

Fig. 24.41: Liver resection for Caroli's disease. (*Courtesy:* Dr Nagaraj Palankar, Assistant Professor, Kasturba Hospital, Manipal)

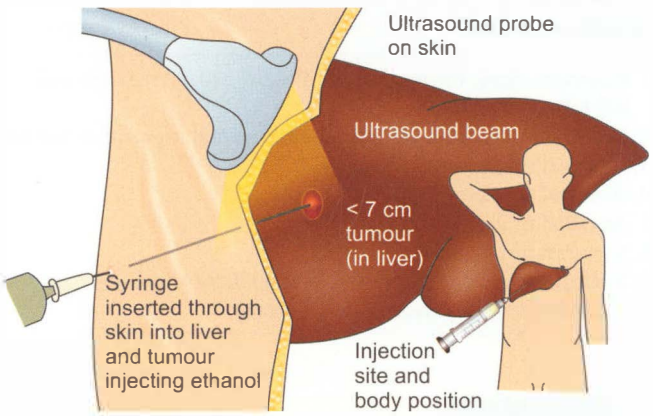


Fig. 24.42: Percutaneous transhepatic ethanol ablation

Embolising agents:

- Gel foam
- Chemotherapeutic agent (like Doxorubicin) with or without Lipiodol
- When radioactive agents are used it is called transarterial radio embolisation (TARE)
- In 'TARE'—implantable radioactive microspheres are delivered into tumour via hepatic artery. Yttrium–90 is most commonly used.

PEARLS OF WISDOM

Lipiodol is derived from poppy seed oil. It is an iodinated ester. It is highly concentrated ester. It is highly concentrated by hepatocytes. The contact time between tumour cells and chemotherapeutic agents is prolonged and it leads to cancer cell death.

4. **Percutaneous ethanol ablation (PEA):** It is cheaper and is a palliative treatment. Tumour less than 3 cm and nodules not more than 3 are candidates. It reduces the tumour size and decreases pain (Fig. 24.42).
   - Tumour necrosis occurs as a result of cellular dehydration denaturation of proteins and occlusion of small tumour vessels.
   - It requires 4–6 sessions under ultrasound guidance with 2–10 ml alcohol injection.
   - Injections are given on an outpatient basis. Easy to give because of the soft consistency of the tumour.

5. **Radiofrequency ablation (RFA)**
   - It is now being tried for inoperable tumours or patients who are not ideal candidates for surgery.
     Indications: Tumour deep in parenchyma, away from hilum and less than 3 cm.

- Electrical energy (500 kHz) is delivered through a 18 G needle inserted under US or CT guidance through the skin into the tumour. In the following few months, the tumour is destroyed and the cells are killed.
- The route of insertion of this needle can be percutaneous, laparoscopic or by open route.
- There should not be any evidence of extrahepatic disease preoperatively or intraoperatively.
  Mode of actions: Cytodestruction of the tumour. In one sitting 6–7 cm of necrosis zone can be achieved for a 5 cm tumour.
- In most cases a tumour can be adequately treated in one session. The procedure can also be repeated. It is done under general anaesthesia.
- It is better to avoid RFA when the tumour is in the hilar plate wherein portal vein and the hepatic arterial branches enter the liver. Damage to these structures and bile duct can occur.

6. **Injection octreotide**, an analogue of somatostatin, has been used in advanced cases. It has shown promising results in decreasing the size of the tumour. It also reduces the pain and discomfort. This is combined with chemotherapy such as gemcitabine and carboplatin. However, the treatment is costly and results are temporary.
7. **Soraferib** is the drug of choice in advanced HCC and with good liver function.

LIVER RESECTIONS

It should be the first choice in HCC, provided there is no extra-hepatic disease or vascular invasion. After getting a negative margin, adequate functioning liver tissue should be present.

Incision

- Aim of the incision is to get adequate exposure
- A transverse abdominal incision in the right upper quadrant with a vertical extension to xyphoid process gives excellent exposure to the liver.

Mobilisation of the liver

- The key point is division of ligaments which are attached to it or which support the liver and suspend the liver.
- The falciform ligament is divided and followed along anterior surface of the liver.
- The left triangular ligament is divided by gentle pull on the left lobe of liver towards right side.
- The right triangular ligament is divided between diaphragm and right lobe of the liver by traction and countertraction.
- Lesser omentum is divided. Thus, separating the liver from stomach.
- Small multiple veins from the inferior surface of the liver join inferior vena cava. These should be divided.
- Last hepatic veins to to be dissected and divided above renal veins.

Hilar dissection

- Cholecystectomy is done by ligating cystic artery and cystic duct. The CBD is then exposed at the free edge of lesser omentum. A sling is applied around CBD to isolate it and gently traction is applied.
- This visualises the common hepatic artery.
- By dissection of common hepatic artery, its right and left branches are dissected out.
- Slings are applied to these arteries, portal vein is exposed. This facilitates freeing of portal vein by dividing lymphatic tissues joining portal vein.
- The congenital anomalies such as aberrant right hepatic artery and accessory left hepatic artery also should be looked for.
- CBD, CHD, left and right hepatic ducts are exposed at the hilum.

Division of the parenchyma

- This requires ligation of artery or branch by 4/0 prolene, transfixation of the bile duct by 4/0 PDS and transfixation of portal vein with 4/0 prolene.
- Once the demarcation appears on the liver, parenchyma is divided by using diathermy.
- Cavitron ultrasonic surgical aspirator (CUSA) is the commonly used dissector for dividing liver parenchyma.
- Vessels and bile duct branches which are exposed are clipped or diathermised.
- Continue dissection till you reach hepatic veins/branches. They are ligated or divided.

Segmental and local resection

- Since each segment (*see* segmental anatomy) carries its own arterial supply, venous drainage and bile drainage, resection can be done with 1 cm margin.
- In a few cases, hilar dissection may not be necessary.

Complications

- Intraoperative bleeding
- Biliary leakage
- Hepatic decompensation
- Ascites due to portal HTN, severing of lymphatics
- Infection

A CASE OF SECONDARIES IN THE LIVER

HISTORY (Table 24.6)

General physical examination

1. Anaemia — It is a common feature of most of the malignancies. Severe pallor may indicate carcinoma stomach or carcinoma colon.

2. Jaundice — Deep jaundice is obstructive in nature. Hence, periampullary carcinoma is likely.

Table 24.6 History taking

Complaint	Explanation
1. Dull-aching and continuous pain in the right hypochondrium of short duration (3–6 months)	Enlarged liver stretches the parietal capsule which is responsible for dull-aching pain
2. Loss of appetite, weakness, asthenia, malaise	Due to destruction of an important metabolic organ
3. Jaundice of a few days' duration, is mild, usually nonprogressive and is not associated with pruritus (hepatocellular variety)	By the time jaundice appears, it is late and enough liver tissue is damaged
4. H/o persistent vomiting with or without blood, with loss of appetite	It suggests that the primary may be in the stomach
5. H/o severe backache without jaundice, disturbing the sleep at night	Carcinoma body and tail of pancreas infiltrating retroperitoneal nerve plexuses
6. H/o jaundice first and associated with itching	Periampullary carcinoma
7. H/o constipation, bleeding per rectum	Primary may be in the colon or rectum
8. H/o previous surgery such as mastectomy	Probably primary is carcinoma of the breast
9. Amputation of toes or leg or wide excision of mole	Malignant melanoma is the likely cause

3. Bilateral pedal oedema	Inferior vena caval obstruction due to enlarged liver.
4. Spine tenderness	Metastasis from breast, prostate, bronchus, kidney, etc.
5. Absent testis in scrotum	Probably seminoma arising from undescended testis.

Abdominal examination

I. Criteria of secondaries in the liver

1. Both lobes are enlarged
2. Lower border is sharp
3. Surface is nodular
4. Hard in consistency
5. Umbilication refers to central necrosis in a nodule but is difficult to appreciate clinically.

II. Evidence of primary

- **Example 1:** Secondaries in the liver from carcinoma stomach. A distinct stomach mass may be felt (Figs 24.43 and 24.44).
- **Example 2:** Secondaries in the liver from periampullary carcinoma. A palpable gall bladder gives clue to the diagnosis (Fig. 24.45).
- **Example 3:** Secondaries in the liver from carcinoma colon or rectum. H/o bleeding per rectum with or without mass. Diagnosis is made by per rectal examination (Fig. 24.46).
- **Example 4:** Secondaries in the liver from seminoma testis. Many times, deep seated seminoma arising from undescended testis cannot be felt per abdomen.
- **Example 5:** Secondaries in the liver from malignant melanoma of foot or leg. Evidence of amputation with enlarged inguinal nodes is usually present (Fig. 24.47).

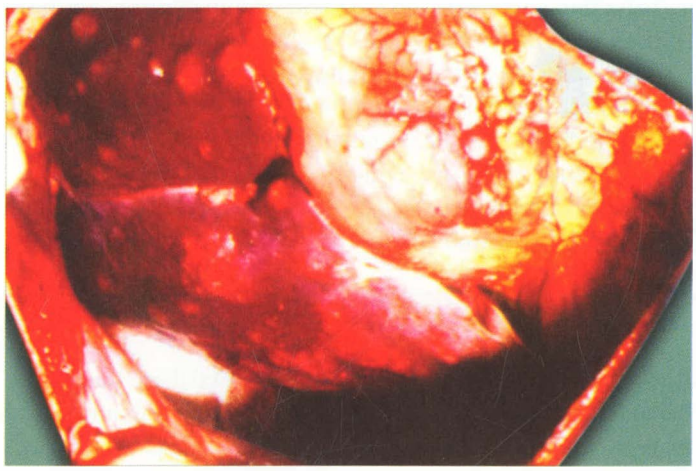


Fig. 24.43: Nodular liver with the stomach mass—stomach is the commonest site of primary

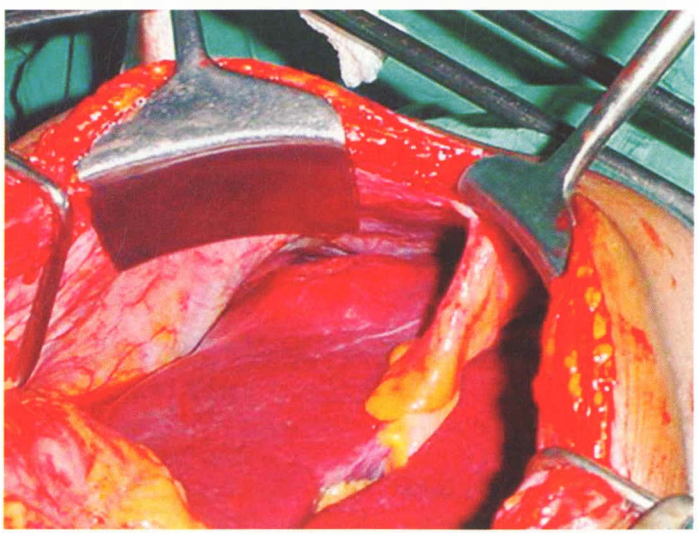


Fig. 24.44: Classical secondary in the liver with umbilication from carcinoma colon. However, it is impossible to appreciate it clinically

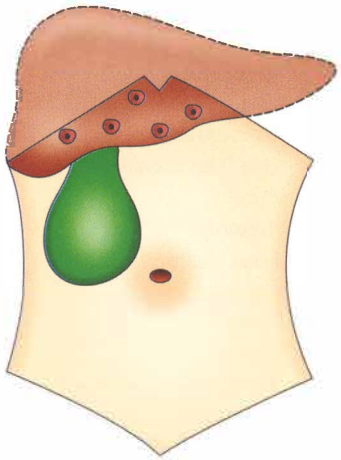


Fig. 24.45: Nodular liver with palpable gall bladder—carcinoma head of the pancreas

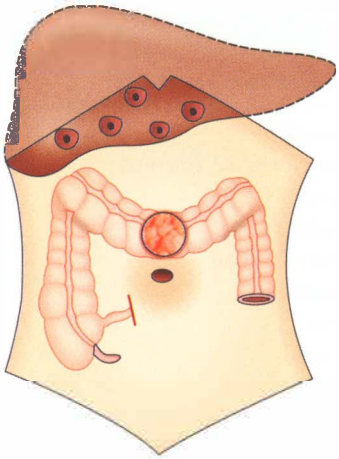


Fig. 24.46: Nodular liver in a case of carcinoma colon—not uncommon to find this

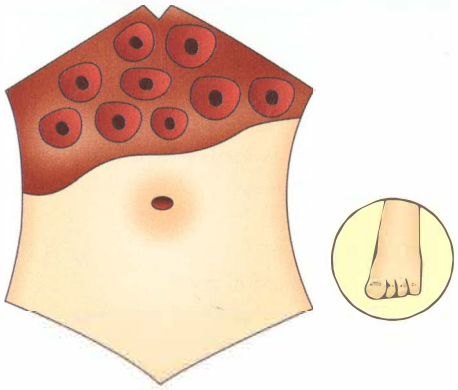


Fig. 24.47: Secondaries in the liver from melanoma—bulky liver with massive hepatomegaly. Note the missing toe

When there are massive secondaries in the liver and if you suspect melanoma—look for
1. Artificial eyeball
2. Missing toes
3. Amputated leg
4. Ulcerated pigmented anal canal
5. A surgical lesion which is erased
6. Wide excision and grafted site
7. Mastectomy scar
8. Laparotomy scar

CLINICAL NOTES

A 38-year-old lady came to the hospital with pain in the right hypochondrium. On examination, firm to hard nodular liver was palpable up to umbilicus. Before investigations, the treating doctor told the husband and relatives that she has secondaries in liver and she may not live for 6 months. 5 years later, she came to the hospital. What she was having was ***Polycystic Disease of Liver!!***— *(Table 24.7)*

Investigations[1]

They depend upon the type of primary which could be detected in a clinical examination. However, when the primary could not be found out, following investigations may have to be done.

1. **Upper gastrointestinal scopy and biopsy:** To detect a growth in the oesophagus or stomach.
2. **Sigmoidoscopy/colonoscopy:** To rule out carcinoma rectum and colon (melanoma of the anal canal).
3. **Abdominal ultrasound**
   - It can diagnose secondaries in the liver which can be hypo- or hyperechoeic and multiple with normal hepatic parenchyma in between.
   - It can detect a carcinomatous growth in the periampullary region.
   - It can detect enlarged lymph nodes—portal, coeliac, para-aortic, etc.
   - It can detect deep seated testicular tumour arising from the testis.

   In spite of these investigations, if the primary cannot be detected, other investigations may have to be done such as bronchoscopy for bronchogenic carcinoma, acid phosphatase for carcinoma prostate or computed tomography (CT scan), etc.
4. High quality CT scan triple phase with IV and oral contrast medium provides actual assessment of the liver disease (decision of hepatectomy) and other pathology in the abdomen.
5. **FDG-PET:** 14-fluorodeoxyglucose positive emission tomography is used to stage the disease, to detect extrahepatic disease.
6. **Liver biopsy:** Laparoscopy and tissue diagnosis is a must if primary cannot be detected. **Lymphoma, carcinoid tumours and metastatic neuroendocrinal tumours have better prognosis.**

Treatment

I. Surgery

In majority of the cases, treatment is only palliative aiming at relieving symptoms. The following are a few examples:

1. Carcinoma stomach with secondaries—palliative GJ, if vomiting is present.
2. Periampullary carcinoma with secondaries—triple bypass to relieve jaundice.
3. In cases of colonic cancer and carcinoid tumour, the primary can be resected because these primaries are slow growing and carry good prognosis. If metastasis is to one lobe of liver, hepatectomy can also be done.

[1]It is important to prove the diagnosis by FNAC or laparoscopic biopsy because if it is neuroendocrinal in origin or is a carcinoid, patient can live a longer life with minimal morbidity.

Table 24.7 Liver mass in the surgical ward

Hepatoma	Secondaries in the liver	Hydatid cyst
1. Not uncommon	Common	Uncommon
2. One lobe is enlarged	Both lobes are enlarged	One lobe is enlarged
3. Consistency is hard or firm	Hard	Firm
4. Surface irregular	Nodular	**Smooth**
5. Short duration	Short duration	Long duration
6. **Tender**	**Nontender**	Nontender
7. General condition poor	Poor	**Good**
8. Vascularity—**thrill and bruit** may be present	Absent	Absent
9. H/o alcohol intake—features of cirrhosis may be present	Symptoms of primary such as loss of appetite, vomiting, backache or another mass may be present	H/o contact with dog may be present

II. Chemotherapy

- When the primary is detected to be in gastrointestinal tract, injection 5-FU can be given. The dosage is 500 mg IV for 5 days/28 days for five cycles.
- When the primary is detected outside the gastrointestinal tract, depending on the nature of the malignancy, appropriate chemotherapy is given (they are discussed in appropriate chapters).

III. Therapeutic embolisation

- A catheter is placed in the hepatic artery and substances such as blood clot or gel foam are injected. They produce thrombosis of hepatic artery resulting in liver necrosis. Thus, liver regresses in size leading to decrease in pain.
- Gel foam is cheap and easy to manipulate. The effect is temporary.
- Autologous blood clot undergoes lysis within 12–24 hours. Hence, it is not ideal.
- Embolisation is an alternative to ligation of hepatic artery.
- Permanent embolising substances include savlon sponge, steel coils, bicrylate, etc.

SOME INTERESTING FEATURES
(Key Boxes 24.13 to 24.16)

Some interesting observations secondaries in the liver

1. Secondaries in the liver from neuroendocrine tumours, carcinoids tumours and colorectal carcinoma have good prognosis.
2. Secondaries in the liver can appear many years after treatment of primary. One example being malignant melanoma of the choroid.
3. It is unusual uncommon for secondaries to develop in a cirrhotic liver.
4. When metastasis appear in the liver from lung cancer, mostly bones, brain, adrenals would have been involved by metastasis, but it is not true/for colorectal cancer. **Often, liver metastasis for colorectal is the only visceral site of metastasis.**
5. More hepatic resections are done for colorectal metastasis than hepatocellular carcinoma.
6. Liver is the most common site of cancer recurrence after a complete resection.
7. Debulking and partial hepatectomy are indicated in metastasis from neuroendocrine tumours.

KEY BOX 24.13

DIFFERENTIAL DIAGNOSIS OF NODULAR LIVER

- Secondaries and hepatoma—hard
- Polycystic disease and hydatid cyst—firm

KEY BOX 24.14

COMMON CAUSES OF SECONDARIES IN THE LIVER

- Carcinoma stomach
- Carcinoma pancreas
- Carcinoma rectum
- Carcinoma colon
- Malignant melanoma
- Testicular tumours

KEY BOX 24.15

CAUSES OF BULKY SECONDARIES

- Malignant melanoma
- Carcinoid tumours
- Colloid carcinoma

KEY BOX 24.16

SPECIAL TYPES OF SECONDARIES

- **Precocious** metastasis—before primary is suspected, e.g. carcinoid, rectal carcinoma
- **Synchronous** metastasis—primary and metastasis detected at the same time, e.g. carcinoma stomach
- **Metachronous** metastasis—metastasis appearing much later than treatment of primary, e.g. melanoma of the choroid.

PORTAL HYPERTENSION

- When portal venous pressure is ***more than 15 mmHg or 20 cm of saline***, it is called portal hypertension.
- Despite high prevalence of varices in patients with cirrhosis, bleeding occurs in only 1/3rd of patients.
- 10–30% of upper GI bleed are due to rupture of varices due to cirrhosis.
- Variceal haemorrhage occurs in 25–35% of patients with cirrhosis.
- Up to 30% of initial bleeds are fatal and as many as 70% of survivors have recurrent bleeds after the first variceal bleed.

Anatomy of portal venous system (Key Box 24.17)

- Splenic vein after receiving inferior mesenteric vein, joins the superior mesenteric vein behind the neck of pancreas and forms the portal vein. It runs in the free edge of the lesser omentum covered by common bile duct.
- It is 6–8 cm in length in its extrahepatic course and divides at the porta hepatis into right and left branches. Rarely, two branches are given on the right side.
- ***Portal vein carries 75% of the blood supply to the liver.***

Left gastric vein enters the portal vein on its anteromedial aspect just cephaloid to the margin of the pancreas. In 25% of cases, left gastric vein joins splenic vein.

KEY BOX 24.17

PORTAL VEIN

Portal vein is 6–8 cm in length
Occurs by confluence of superior mesenteric vein and splenic vein
Runs posterior to common bile duct
Two venous communications between coronary and azygos veins give rise to varices
Approximately, it supplies 75% of total blood flow to the liver
Ligament related to portal vein is hepatoduodenal
Valveless structure
Esophagogastric varices develop if there is portal hypertension
Increased portal pressure results in liver damage and bleeding
Normal pressure is 3–5 mmHg
Remember as **PORTAL VEIN**

Aetiology (Fig. 24.48)

I. Extrahepatic portal hypertension (prehepatic)

1. Splenic vein thrombosis

2. Portal vein thrombosis

It is secondary to umbilical vein sepsis in neonates (omphalitis). In adults, thrombosed portal vein is surrounded by a leash of vessels 'cavernoma'. Portal vein thrombosis can also occur due to carcinoma of the pancreas in elderly patients.

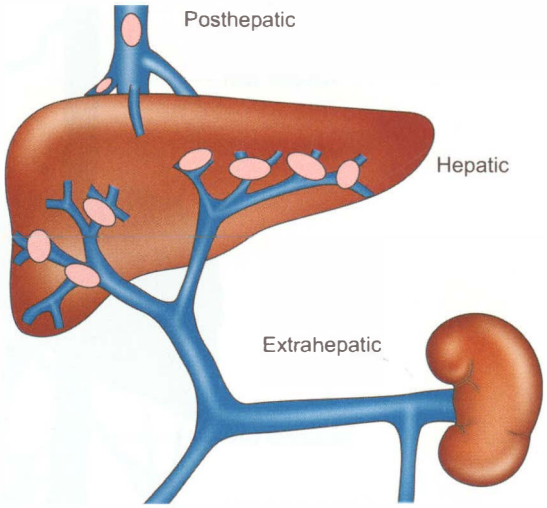


Fig. 24.48: Causes of portal hypertension

3. Portal vein agenesis

Extrahepatic portal hypertension is seen in 20–25% of cases and is common in female children. Liver function is normal in these cases.

II. Intrahepatic portal hypertension

It accounts for nearly 80% of cases.

1. Cirrhosis of the liver

The regenerating nodules compress the portal venous radicles within the liver. Liver function is very poor.

2. Schistosomiasis

Common in Egypt. *Schistosoma mansoni* and *S. japonicum* lay eggs within branches of portal vein and cause hepatic fibrosis resulting in portal hypertension.

III. Posthepatic portal hypertension

These are medical causes:

1. Tricuspid incompetence
2. Constrictive pericarditis
3. Budd-Chiari syndrome

Pathophysiology

As a result of obstruction to portal vein, in an attempt to reduce portal pressure, the normally present insignificant collaterals open up[1]. They become significant and result in development of portosystemic anastomosis.

- Even though, portosystemic shunts develop in many places, their important locations and clinical significance have been outlined in Table 24.8 and Fig. 24.49.

[1]When the main highway is blocked, small insignificant roads become significant, is it not?

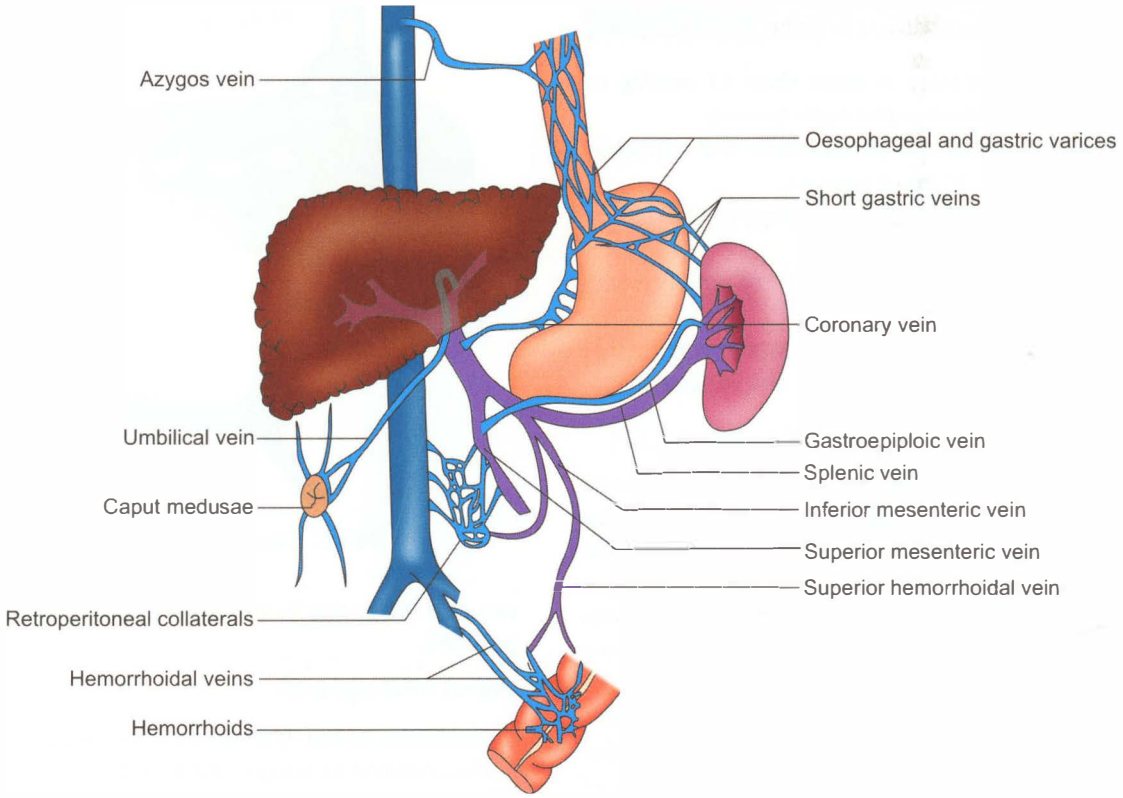


Fig. 24.49: Collaterals

Table 24.8 Anastomosis between the portal and systemic venous system

Site	Portal vessels	Systemic vessels	Effect
1. Lower end of oesophagus	Branches of left gastric vein and short gastric vein	Branches from azygos vein	Oesophageal varices
2. Falciform ligament of liver (paraumbilical vein)	Veins which run in the falciform ligament	Anterior abdominal wall veins	Caput medusae*
3. Lower end of rectum	Superior haemorrhoidal vein	Inferior and middle haemorrhoidal vein	Piles (very rare), rectal varices
4. Retroperitoneum mesenteric veins	Branches of superior and inferior subdiaphragmatic veins	Retroperitoneal veins	Retroperitoneal varices (silent)

*Most often,veins which run in the falciform ligament are obliterated from birth. Hence, it is not commonly seen.

- The lower end of the oesophagus is one of the important areas giving rise to oesophageal varices. These varices rupture resulting in massive haematemesis.
- This is exactly the reason why every attempt at newer modality of treatment, medical or surgical, is aimed at controlling oesophageal varices.

Clinical features (Table 24.9)

Bleeding gastroesophageal varices are one of the common causes of death among cirrhotics patients.

Prediction of variceal haemorrhage

1. **Physical factors:** Elastic properties of vessels, intravariceal and intraluminal pressure, and variceal wall tension are major factors.
2. **Continued alcohol use**
3. **Poor liver function**
4. **Hepatic venous pressure gradient (HVPG)**

- HVPG = (wedged/occluded hepatic venous pressure)—(free hepatic venous pressure). **Normal gradient** is **5 mmHg**. HVPG > 12 mmHg indicates portal hypertension.

Investigations

1. Complete blood picture

- Anaemia is usually present due to bleeding.
- Peripheral smear to rule out hypersplenism—pancytopaenia in the presence of normal bone marrow.

2. Liver function tests

- Serum proteins: Total protein, albumin are low and albumin : globulin ratio is reversed in cirrhosis of the liver.

Table 24.9 Clinical comparison of extrahepatic with intrahepatic portal hypertension

	Extrahepatic	Intrahepatic
1. Age	10–25 years	> 40 years
2. H/o alcohol	Absent	Strongly positive
3. H/o jaundice	Absent	Present
4. Sex	More in females	More in males
5. Splenomegaly	Moderate to big	Mild to moderate
6. Ascites	Absent	Present
7. Liver cell failure	Absent	Present
8. Encephalopathy	Not a feature	It is common

- Enzymes SGOT (serum glutamic oxaloacetic transaminase) and SGPT (serum glutamic pyruvic acid transaminase) are increased in hepatocellular failure.
- Serum bilirubin: Increased levels indicate liver cell failure.
- Prothrombin time, bleeding time and clotting time must be checked before a liver biopsy is done.
- **Liver biopsy** is done provided bleeding profile is normal. However, injection vitamin K 10 mg, IM is given for 3 to 5 days.
- HBsAg serum hepatitis predisposes to cirrhosis of the liver.
- Liver functions are usually normal in extrahepatic portal hypertension.

3. Oesophagogastroduodenoscopy (OGD)

- To confirm oesophageal varices which appear as a bluish red longitudinal column in the lower end of the oesophagus. When they extend to the stomach, they are called gastric varices (Figs 24.50 to 24.52).
- Cherry red spots indicate impending rupture
- Grading of varices (Key Box 24.18).

4. Splenoportovenography (SPV)

A fine lumbar puncture needle is passed through the 9th space in the left midaxillary line into the spleen and a radiopaque dye injected into the splenic pulp which fills up the entire portal system.

Requirement and precautions

- BT, CT, PT should be normal.
- Prophylactic antibiotics are given to avoid pyaemia.

KEY BOX 24.18

ENDOSCOPIC GRADING OF VARICES BASED ON VARIX HEIGHT AT 2 CM FROM OG JUNCTION

Grade	Bite width 1.7 mm forceps = 5 mm
1+	One-fourth bite width
2+	One-half bite width
3+	Three-fourths bite width
4+	One or more bite width(s)

Fig. 24.50: Oesophageal varices

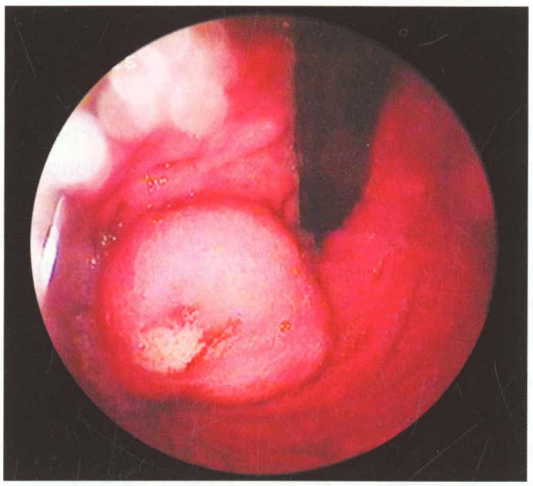


Fig. 24.51: Gastric varices

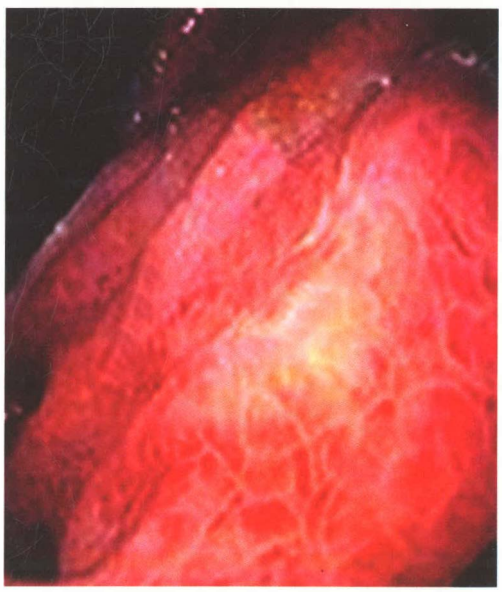


Fig. 24.52: Gastropathy

Fig. 24.53: SPV showing dilated splenic vein

Uses (Fig. 24.53)

- To know the anatomical configuration of portal system, so that a proper shunt operation can be done.
- To locate the exact site of obstruction.
- To assess the diameter of splenic vein. When it is more than 1 cm, it is an indication for splenorenal shunt (lieno-renal shunt).
- If the needle is connected to a manometer, splenic pulp pressure can be recorded.

5. Ultrasonography

To assess the nature of the liver (**cirrhosis**), to know the portal vein, portal cavernoma, splenic vein diameter, etc. It has become a valuable investigation today.

Causes of rupture of varices

- Variceal bleeding can occur even without any specific cause. The bleeding can be very minor in the form of occult blood in the stools to massive bleeding. Following hypothesis has been postulated for massive bleeding from varices.

  Eruptive theory: Due to increased intravariceal pressure.

  Erosive theory: Erosion of mucosa over the varices.
- Bleeding manifests as haematemesis or malaena. Haemodynamic instability is common.

Treatment of portal hypertension—guidelines

I. Primary prophylaxis
II. Secondary prophylaxis
III. Acute variceal haemorrhage

I. Primary prophylaxis (before bleeding)

This is aimed at reducing portal pressure and consequently intravariceal pressure.

A. Pharmacotherapy: Drugs used are **nonselective β-blockers** alone (propranolol) or in combination with **isosorbide mononitrate**.

- Reducing portal pressure by at least 20% or HVPG < 1 mmHg is associated with significant protection against bleeding.
- The dose of β-blocker is titrated to maintain heart rate around 55 beats/min or reducing by 25% from baseline rate.

B. Endoscopic therapy: Endotherapy

- **Endoscopic band ligation** of varices is better than sclerotherapy. It is very effective and associated with less side-effects.
- Indicated in patients **who do not tolerate β-blockers** and varices which are large and are associated with red wheal markings.

II. Secondary prophylaxis

- It means prevention of rebleeding following acute variceal haemorrhage.
- The different forms of treatment and their success depends upon various criteria. The most popular of these is the Modified Child's criteria (Table 24.10). Serum albumin, bilirubin, ascites, encephalopathy and prothrombin time are included under this criteria.

Table 24.10 Modified Child's criteria Grade A = 5–6 points, Grade B = 7–9 points, Grade C = above 10 points

	Points awarded for abnormality		
	1	2	3
1. Albumin g/L	35	28–35	< 28
2. Bilirubin mol/L	15–30	30–45	> 45
3. Ascites	Absent	Easily controlled	Not controlled
4. Encephalopathy	Absent	Mild	Severe
5. Prothrombin time (in seconds)	1–4	4–6	Above 6

III. Treatment of massive bleeding from varices

1. General measures

- Admission, hospitalisation, preferably in an intensive care unit, elevation of foot end of the bed to increase venous return.
- Intravenous cannulation or a venous cut down and replacement of fluid till blood is ready.
- Blood transfusion.

2. Measures to prevent encephalopathy

- Ryle's tube is introduced and stomach wash is given with ice-cold saline.

- Bowel wash is given to decrease blood from gut, to decrease ammonia and uric acid levels so as to prevent encephalopathy.
- Oral neomycin 1 g, 6th hourly as intestinal antiseptic.
- Oral lactulose 30 ml, 8th hourly is given. It also helps in decreasing encephalopathy.

3. Pharmacotherapy for variceal bleeding

- **Injection Vasopressin,** 20 units in 200 ml of saline is given intravenously over a period of 20 minutes. This dose avoids coronary and peripheral vasoconstriction. By causing powerful splanchnic vasoconstriction, it decreases the blood flow to the varices and reduces bleeding in about 50% patients.
- **Injection Somatostatin** 250 µg IV bolus followed by an infusion of 50 µg/h. It is very effective and has no side-effects. However, it is costly.
- **Injection Octreotide** and **Terlipressin** are preferred because of their long half-life. They act by producing splanchnic vasoconstriction. Thus, they decrease the portal pressure. Once started, they have to be continued for 2–5 days. Once bleeding stops, the varices are treated with **endotherapy** to prevent rebleeding.
- **Metoclopramide** 20 mg IV arrests the bleeding by constricting gastro-oesophageal sphincter.

4. Endotherapy—types

A. Variceal banding (Key Box 24.19 and Fig. 24.54)

- Today, variceal banding has become the gold standard for oesophageal varices because of their efficiency in controlling the bleeding and less chances of complication.
- In 10% of the cases, rebleeding can occur.

B. Sclerotherapy (Key Box 24.20 and Fig. 24.55)

- 2% ethanolamine oleate or sodium tetradecyl sulphate (STD) is used, 3–5 ml into each varix **(intravariceal)** or 0.5 ml into the side of varices **(paravariceal)**
- Bleeding can be controlled in about 80–90% of patients. If the bleeding continues, repeat injections can be given.
- If the bleeding continues in spite of the above measures, balloon tamponade is done.

Disadvantages of endoscopic sclerotherapy

1. Gastric varices cannot be managed by sclerotherapy.
2. Multiple oesophageal ulcers can develop at the injection site.
3. 10–20% of the patients develop low grade fever.
4. It can precipitate massive bleeding.
5. It can cause mediastinitis, left pleural effusion.
6. Chances of rebleeding are high. Hence, repeated injections are necessary.

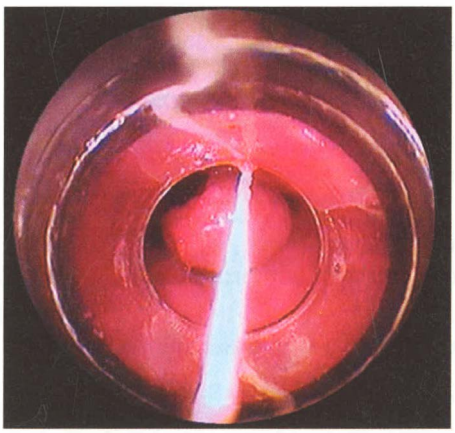


Fig. 24.54: Endoscopic banding

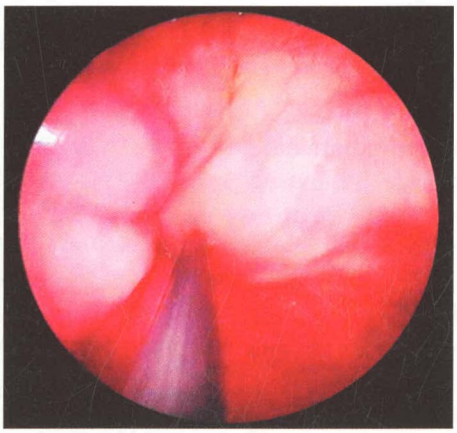


Fig. 24.55: Endoscopic sclerotherapy

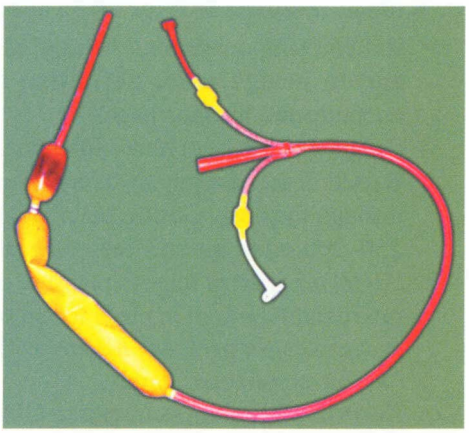


Fig. 24.56: Balloon tamponade

(*Courtesy:* Dr Filipe Alvares, Medical Gastroenterologist, KMC, Manipal)

KEY BOX 24.19

VARICEAL BANDING

Advantages

- Controls bleeding in 80–90% of cases
- Less rebleeding rates
- Easy
- Less risk of complication such as perforation or stricture

Disadvantage

- Costly

KEY BOX 24.20

SCLEROTHERAPY

Advantages

- Cheap
- Easy
- More commonly available

Disadvantages

- Complications such as perforation or stricture and massive bleeding can occur
- Higher rebleeding rates.

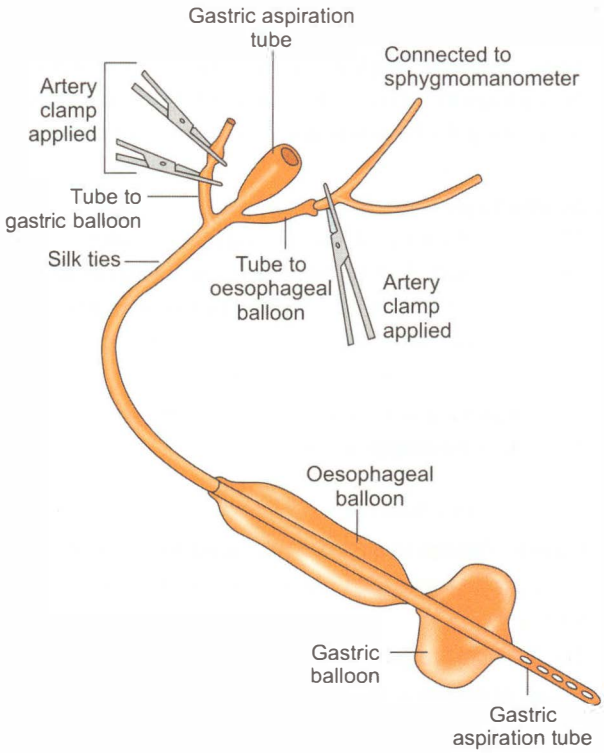


Fig. 24.57: Sengstaken tube sketch

C. Balloon tamponade (Figs 24.56 and 24.57)

- Balloon tamponade must be used as a **rescue procedure** and to bridge more definitive therapy in case of uncontrolled haemorrhage.
- Sengstaken's tube or its modification, the **Minnesota tube** is used. It acts directly by internal tamponade. It is passed through the nose. Gastric balloon is distended with up to 250–300 ml of air and oesophageal balloon with about 50–60 ml of air or to get a pressure of 20–30 mmHg. The tube cannot be kept in place for more than 12 to 24 hours as it can cause **pressure necrosis**. The method is reliable only in experienced hands.
- Recurrent bleeding is common after decompression.

D. TIPSS: Transjugular Intrahepatic Portosystemic Stent Shunt (Fig. 24.58)

- Consists of vascular placement of expandable metal stent across the tract created between hepatic vein and major branch of portal system.
- TIPSS helps a small group of patients (5 to 10%) who have refractory bleeding.
- Low morbidity and mortality.
- Incidence of encephalopathy is similar to surgical shunts. (*See* Figs 24.59 for management of acute oesophageal variceal bleeding.)

5. Surgery

- It is of two types—devascularisation procedures and shunt procedures.

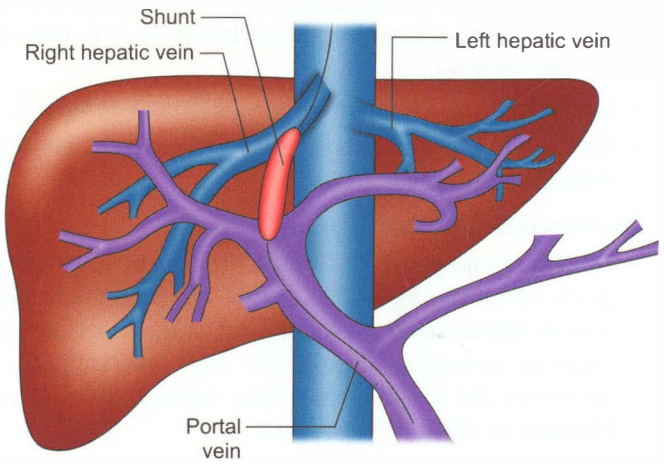


Fig. 24.58: TIPSS

PEARLS OF WISDOM

Devascularisation procedures are aimed at reducing the pressure in the varices by disrupting the blood supply and shunt procedures are aimed at reducing portal pressure.

A. Devascularisation procedures

1. Oesophageal transection

- It is done through left thoracoabdominal route called 'Milnes-Walker operation'.
- During the transection, varices are disconnected, bleeders are ligated or under-run, followed by oesophagogastric anastomosis.

2. Gastric transection of Tanner

In this operation the division is made in the stomach through abdominal route.

3. Sugiura and Futugawa operation

It is an extensive devascularisation procedure.

Aim

To interrupt the intramural and submucosal veins.

Procedure (Figs 24.60 and 24.61)

In this operation, the following procedures are done:

- Splenectomy
- Devascularisation of the greater curvature of the stomach and devascularisation of lesser curvature of the stomach as in highly selective vagotomy.
- Oesophagogastric transection and suturing by using **EEA** (**E**nd to **E**nd **A**nastomosis) staplers. It is a major surgery. Mortality is around 10–15%. It is the most effective devascularisation.

B. Portosystemic shunt procedures

Indications

1. Continuing variceal bleeding in spite of sclerotherapy wherein the general condition of the patient is good (Child's A, Child's B).

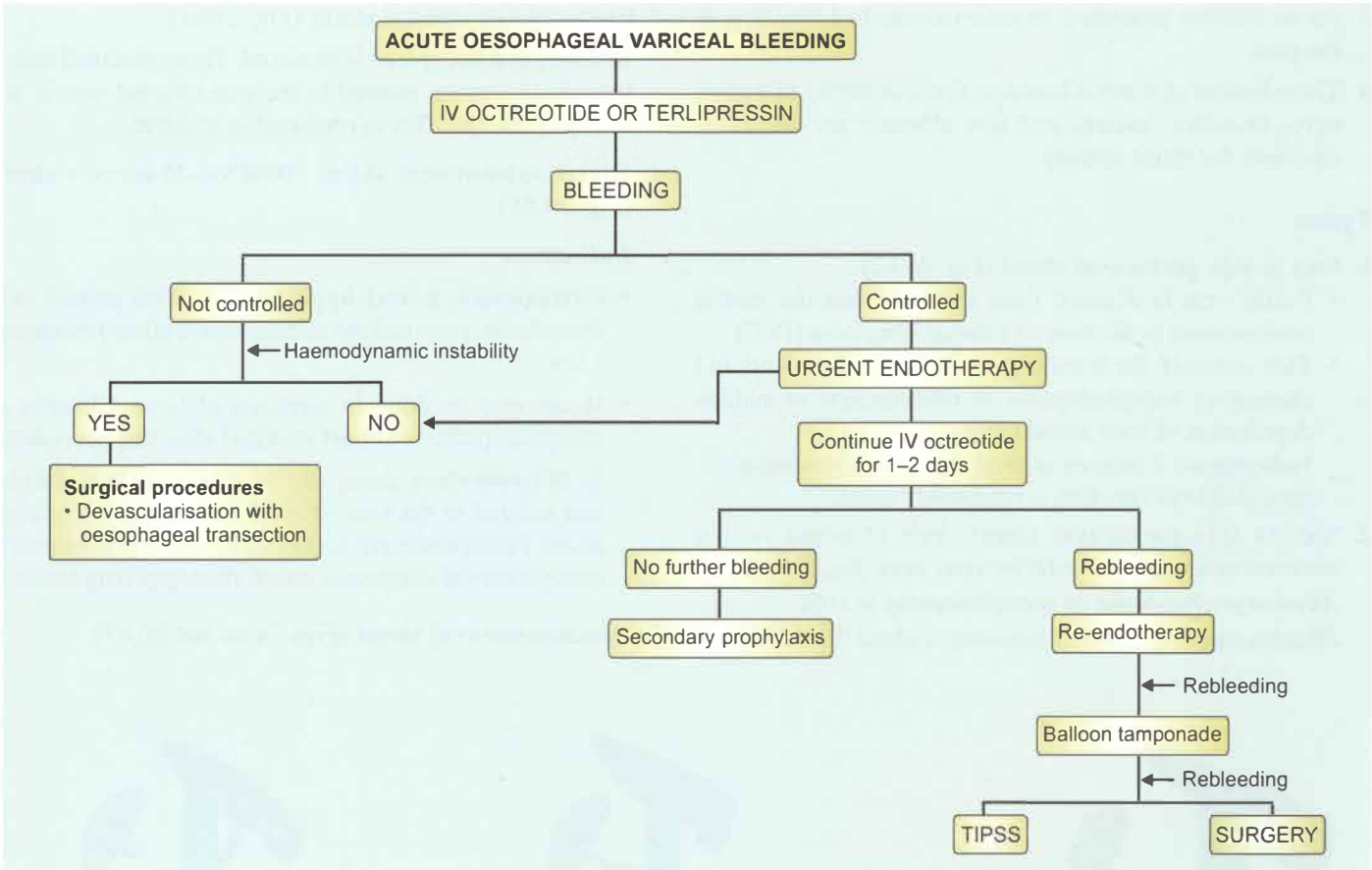


Fig. 24.59: Algorithm showing management of bleeding varices

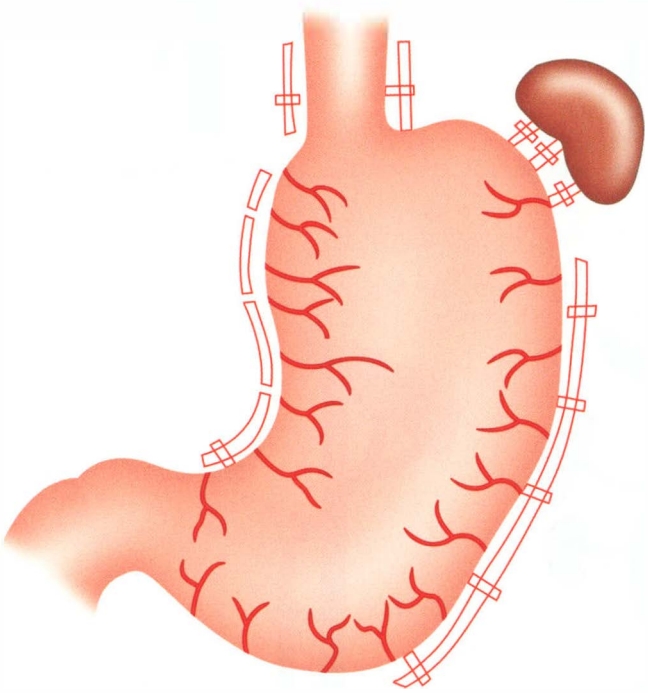


Fig. 24.60: Sugiura and Futugawa operation (splenectomy with devascularisation). Ideally done through thoracoabdominal incision

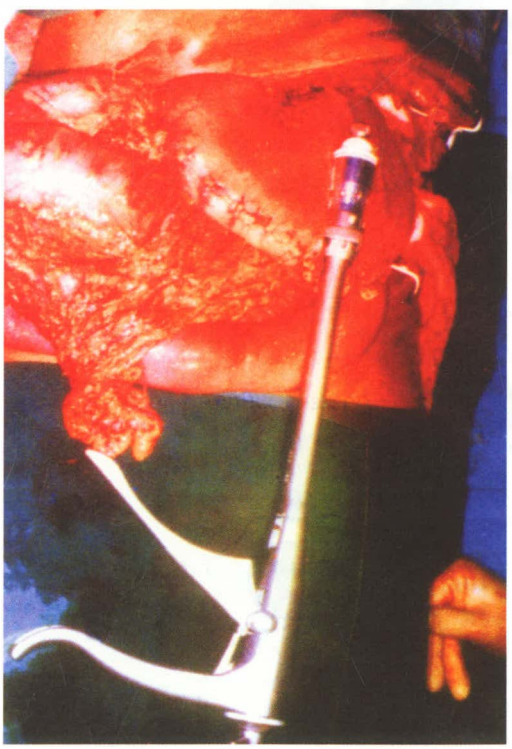


Fig. 24.61: Usage of EEA staplers for devascularisation procedure (*Courtesy:* Prof U Santhosh Pai, KMC, Manipal)

2. As an elective procedure in patients who had bleeding in the past.
3. The selection of shunt is based on the availability of a good vein. Jaundice, ascites and low albumin are contraindications for shunt surgery.

Types

1. End to side portocaval shunt (Fig. 24.62)

- Portal vein is divided from the liver and the end is anastomosed to the side of inferior vena cava (IVC).
- This controls the bleeding in 90–95% of patients but chances of encephalopathy is 30% because of sudden deprivation of liver blood supply.

Indications: Cirrhosis of liver due to schistosomiasis, provided liver function is reasonably good.

2. Side to side portocaval shunt: Side of portal vein is anastomosed to side of inferior vena cava (Fig. 24.63).

Advantage: Incidence of encephalopathy is 10%.

Disadvantage: Control of bleeding is about 50–70%.

3. Proximal splenorenal shunt (Fig. 24.64)

In this operation, spleen is removed. Then, proximal end of the splenic vein is sutured to the side of renal vein in the retroperitoneum. This is preferred in children.

4. Distal splenorenal shunt (DSRS)—Warren's shunt (Fig. 24.65)

Indications

- Extrahepatic portal hypertension with portal vein thrombosis, provided the splenic vein is dilated more than 1 cm.
- It can also be done in cirrhosis of liver. Chances of encephalopathy is almost minimal after this procedure.

In this operation, distal end of splenic vein is divided and sutured to the side of renal vein in the retroperitoneum. Postoperatively, ascites can occur due to extensive retroperitoneal dissection which damages lymphatics.

5. Mesentericocaval shunt (Figs 24.66 and 26.67)

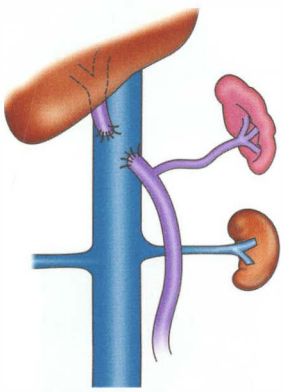


Fig. 24.62: End to side portocaval shunt

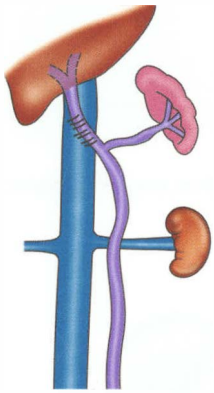


Fig. 24.63: Side to side portocaval shunt

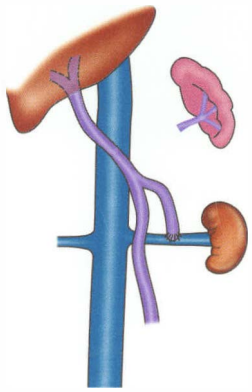


Fig. 24.64: Proximal splenorenal shunt

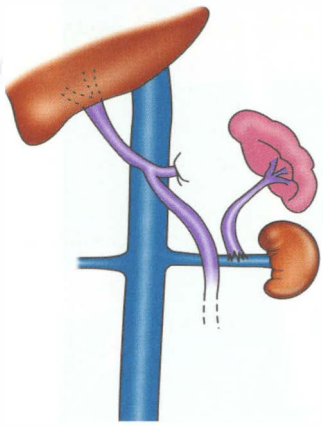


Fig. 24.65: Distal splenorenal shunt (DSRS)

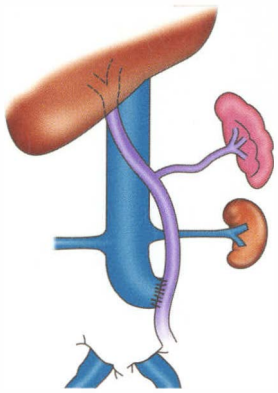


Fig. 24.66 : Mesentericocaval shunt

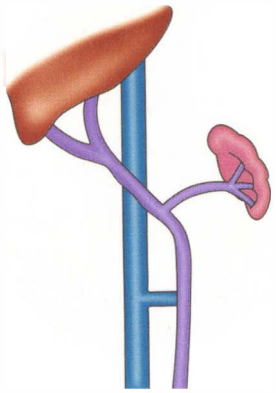


Fig. 24.67: Mesentericocaval shunt—H-graft

Indications

1. Splenic vein is not dilated or thrombosed.
2. Portal vein is thrombosed

Procedure

- **The inferior vena cava is divided** and its proximal end is sutured to the side of superior mesenteric vein (SMV).
- Otherwise, a dacron graft can be placed between IVC and SMV. This is described as mesentericocaval H-graft or jump graft.
- In children, IVC can be divided without fear of pedal oedema.

Shunt procedures have to be undertaken only by an experienced surgical gastroenterologist in an institution. These patients have to be carefully monitored in the postoperative period for possible blockage of shunt.

CONTROL OF ASCITES IN PORTAL HYPERTENSION

- Ascites in portal hypertension is a slow, insidious accumulation of free peritoneal fluid, an occurrence in advanced liver disease (Key Box 24.21).
- It can be troublesome and refractory to commonly available treatment.
- Diagnosis is confirmed by ultrasound and aspiration.
- When there is a doubt regarding the diagnosis, **laparoscopy** is advisable which not only detects and confirms cirrhosis but also rules out other intra-abdominal pathologies (malignancy, tuberculosis, etc.).

Treatment (Key Box 24.22)

1. **Diuretics**
   K^+ sparing diuretics such as spironolactone or frusemide can be given. However, hyponatraemia and hypokalaemia should be monitored.
2. **Restriction of salt intake:** 20 mg/day is advised.
3. **Peritoneovenous shunt: Le Veen shunt**
   - Le Veen shunt is done by using a silastic tube to insert into ascites. It is then tunnelled subcutaneously to the neck and inserted under vision into internal jugular veins.
   - **Mode of action:** Ascitic fluid is drawn into systemic circulation with each respiration because of the presence of a one-way valve.

KEY BOX 24.21

CAUSES OF ASCITES IN PORTAL HYPERTENSION

- Reduction in plasma albumin
- Central hypovolaemia
- Retention of salt and water by kidneys
- Portal hypertension

Complications

- Occlusion
- Displacement
- Infection

KEY BOX 24.22

TREATMENT

Salt restriction
Abdominal paracentesis
Liver transplantation
TIPSS
Intake of diuretics
SHunt—peritoneovenous
Remember as **SALTISH**

MISCELLANEOUS

TEN COMMANDMENTS OF BLEEDING VARICES

1. All cases of cirrhosis **should undergo** upper gastrointestinal endoscopy to rule out varices.
2. Almost all cases of varices **should receive propranolol** prophylaxis. Propranolol prophylaxis reduces the risk of bleeding in varices by at least 10%.
3. Acutely bleeding oesophageal varices **should be first** treated with octreotide and endoscopic treatment.
4. Endoscopic sclerotherapy, and variceal banding **should be tried** in all patients with variceal bleeding. Banding is safer than sclerotherapy.
5. One should **consider TIPSS** if available, when pharmacotherapy and endoscopic therapy fail.
6. One should **consider Sengstaken-Blakemore** tube if endoscopic facilities and TIPSS are not available although it is associated with a high rate of recurrent bleeding.
7. Inexperienced surgeon **should consider oesophageal transection** to tide over bleeding varices resistant to octreotide and endoscopic treatment.
8. One **should consider Child's score** before planning measures to prevent rebleeding.
9. Surgical **shunts should be considered for Child's A and B** category.
10. In **Child's C category, one should consider TIPSS followed by liver transplant**.

PORTAL HYPERTENSIVE GASTROPATHY

- Due to increased portal pressure and collaterals, vascular dilatation and ectasia occur in the stomach.

- Endoscopically they appear pink speckled with red mosaic like pattern in the gastric mucosa.
- Lesions are diffuse (unlike GAVE—*see* page 485)
- They are responsible for about 3.5% of cases of upper GI bleeds and 15% in portal hypertension cases.
- They can present as acute bleeding with needs to be managed conservatively.
- Chronic bleeds present as anaemia.
- Propranolol is used to reduce portal pressure which in turn reduces bleeding.

PORTAL BILIOPATHY (PSEUDOSCLEROSING CHOLANGITIS)

- It refers to bile duct changes due to portal hypertension.
- More common in extrahepatic portal hypertension.
- Portal cavernosa, ischaemic injury to the bile duct are the causes. Infection is also one of the causes.
- Changes in the bile duct include strictures and dilatation of both extra and intrahepatic bile ducts secondary to varices encircling CBD and gall bladder wall. Choledocholithiasis is common.
- Can present with bile duct obstruction, cholangitis and bleeding into the CBD.
- Prolonged obstruction leads to secondary biliary cirrhosis.
- Investigation: U/S, CT scan, colour Doppler to image varices
- Treatments: Endoscopic sphincterotomy, clear the CBD, stenting.
- Portosystemic shunt in failed endoscopic extraction of stone.

BUDD-CHIARI SYNDROME

It occurs due to obstruction to the hepatic vein.

Causes

1. Congenital: It is a potentially curable condition where in a web exists in the suprahepatic portion of inferior vena cava.
2. Clotting disorders—polycythaemia
3. Contraceptive pills
4. Cancerous infiltration of hepatic veins (hepatocellular carcinoma).
5. Crotalaria, a plant extract used in the tea.

Clinical features

1. **Acute form** is dangerous, resulting in rapid enlargement of liver, severe abdominal pain, vomiting and hypotension.
2. **Chronic form** resembles cirrhosis
   - Hepatomegaly—firm, can be irregular, dilated veins over the abdominal wall. Bilateral pedal oedema which can be irritating, signs of liver cell failure are seen. Haematemesis occurs later.

Treatment

In congenital cases, if web can be proved by venacavography, it can be excised by transatrial meatotomy. However, in general, prognosis is very poor. Peritoneovenous shunt has to be considered as an alternate method to drain the ascitic fluid into one of the veins in the neck (internal jugular vein).

ROLE OF OCTREOTIDE IN SURGERY

1. **Variceal bleeding**
   - **Acute**—50 µg IV stat followed by 50 µg/h infusion for 120 hours.
   - Long-term **prophylaxis** is 100 µg s/c tds for 15 days.
2. **Pancreatic disorders**
   - ERCP-induced pancreatitis
   - Acute pancreatitis
   - Relapsing pancreatitis
   - Chronic pancreatitis
   - Pancreatic surgery—100 µg s/c 1 h prior to surgery and to continue 100 µg tds for 7 days.
3. **Bowel dysfunction**—AIDS-related diarrhoea, short bowel syndrome, dumping syndrome, chemotherapy-induced diarrhoea.
4. **Gastrointestinal fistulae**
5. **Carcinoid and other neuroendocrine tumours.**
6. **Advanced malignancies:** Hepatocellular carcinoma, metastatic breast cancer, colonic cancer, pancreatic cancer.
7. **Acromegaly**

LIVER TRANSPLANTATION

Indications

- Decompensated cirrhosis: Chronic hepatitis C, primary biliary cirrhosis, etc.
- Acute fulminant hepatic failure (FHF): Causes are para-cetamol overdosage, acute hepatitis B infection, Wilson's disease, drugs.

Signs and symptoms of decompensated liver

- Hepatic encephalopathy
- Ascites
- Spontaneous bacterial peritonitis
- Hepatorenal syndrome
- Hyperbilirubinaemia, coagulopathy

Types

1. **Living donor transplant:** Easily available, no waiting period but potential risk to donor (normal person) is present.
2. **Cadaveric:** Preserving organ by hypothermia (Key Box 24.23)
   - Waiting period
   - No risk to donor
   - To be done within 12 hours, ideally within 6 hours.

KEY BOX 24.23

ORGAN PRESERVATION

- Hypothermia and by cold storage solutions
- University of Wisconsin solution is commonly used.
  It contains:
  – Lactobionate: Prevents intracellular swelling
  – Hydroxyethyl starch: Decreased interstitial oedema, decreased hypothermia—induced damage to cells.

Type of liver transplantation

- Partial hepatectomy: Usually right lobe (60% of total liver)
- Split liver transplant: Divide into two functioning units and transplant into two patients.

Procedure

- Mobilise the diseased liver and isolate vena cava, portal vein, hepatic artery and bile duct.
- Remove the diseased liver
- Donor liver is transplanted and anastomosis of various structures done.

Complications

- Thrombosis of hepatic artery
- Thrombosis of portal vein
- Bile leak
- Nonfunctioning of hepatic allograft
- Infections

*More details of liver transplantation are given in page 1109.

HAEMOBILIA

Definition

Bleeding from liver into the biliary tree. There is usually a **communication between a blood vessel and the bile duct** or any other part of the biliary tree.

- Accidental trauma
- Malignant liver disease
- Portal hypertension
- Percutaneous diagnostic and therapeutic procedures
- Vascular disease of hepatic artery
- Gallstones may get impacted, erode the hepatic artery and cause life-threatening haemobilia.

Clinical features

- **Triad of Sandblom**—jaundice, pain and melaena
- Pain abdomen—colicky
- Obstructive jaundice
- Haematemesis or melaena

Investigation

LFT, USG abdomen, selective arteriography, upper GI endoscopy.

Treatment

- Selective arterial embolisation (angiographic, under fluoroscopic guidance)
- Blood transfusions and antibiotics to prevent cholangitis.
- Causes of hypoalbuminaemia are mentioned in Key Box 24.24.

KEY BOX 24.24

CAUSES OF HYPOALBUMINAEMIA

Sepsis
Enteropathy—protein-losing
Poor nutrition—malnutrition
Syndrome—nephrotic
Injury—trauma
Sick Liver—Chronic Liver Disease
Remember as **SEPSIS**

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All the topics have been updated
- New photographs and key boxes have been added
- Liver transplantation which is being increasingly done in India has been included under transplantation in Chapter 49
- Cystic diseases have been added
- Haemobilia is added

MULTIPLE CHOICE QUESTIONS

1. Which of the following statements is false about hepatic encephalopathy?
A. It is caused by ammonia
B. Precipitated by sedatives
C. Not precipitated by bleeding into gastrointestinal tract
D. Precipitated by diuretics

2. About hepatorenal syndrome following are true *except:*
A. It can occur in sepsis
B. Most of them have advanced liver disease
C. It is associated with profuse intrarenal vasodilatation
D. Orthotopic liver transplantation can reverse hepatorenal syndrome

3. Which of the following is not the cause for over-production of bilirubin?
A. Total parenteral nutrition
B. Haemolytic anaemias
C. Blood transfusion
D. Gilbert's syndrome

4. Triad of Sandblom include the following *except*:
A. Colicky abdominal pain
B. Obstructive jaundice
C. Haematemesis or melaena
D. Sepsis

5. The following are indications for percutaneous method for drainage of pyogenic liver abscess *except*:
A. Superficial abscesses
B. Abscess with no intra-abdominal pathology
C. Abscesses of unknown aetiology
D. Multiple abscesses

6. Very tender liver is a feature of the following *except*:
A. Secondaries in the liver
B. Amoebic liver abscess
C. Hepatoma
D. Congestive cardiac failure

7. A differentiating feature of amoebic liver abscess from acute cholecystitis is:
A. Intercostal tenderness
B. Abdominal tenderness
C. Guarding
D. Fever

8. The drug of choice for the treatment of amoebic liver abscess is:
A. Penicillin B. Gentamicin
C. Metronidazole D. Cotrimoxazole

9. The following are scolicidal agents *except*:
A. Hypertonic saline
B. Chlorhexidine
C. Sodium hypochlorite
D. Hydroxyethyl starch

10. Indications for PAIR in the treatment of hydatid cys includes:
A. Relapse following surgery
B. Multiple septal divisions
C. Communicating cysts
D. Dead or inactive cysts

11. The following feature is true about *Echinococcus multilocularis except*:
A. Alveolar echinococcosis
B. Causes malignancy
C. Poorly demarcated honeycombed cystic lesion
D. Infiltrates liver and invades vascular system

12. A 65-year-old patient presents with the following: Serum bilirubin: 4 mg%, serum albumin: 1.8 mg%, INR: 2. He has gross ascites and is drowsy. What Child-Pugh Class does he belong to?
A. A B. B
C. C D. D

13. The following are features of polycystic liver disease *except*:
A. They are usually asymptomatic
B. The cysts do not produce liver cell failure
C. They are acquired
D. Cysts also occur in kidneys and pancreas

14. Which of the following is the commonest benign tumour of the liver?
A. Haemangioma
B. Adenoma
C. Focal nodular hyperplasia
D. Hepatoma

15. For purposes of hepatic resection, the liver can be divided into _____ number of segments.
A. 2 B. 4
C. 6 D. 8

16. The main portal fissure is also called:
A. Cantlie's line
B. Murphy's line
C. Spigelian line
D. Macewan's line

17. The following conditions are associated with increased risk of hepatocellular carcinoma *except:*
A. Hepatitis B carrier state
B. Cirrhosis
C. Malignant hydatid cyst
D. Aflatoxin consumption

18. Up to _____ segments of the liver can be resected safely without the danger of hepatic failure provided rest of the liver is normal.
A. 2 B. 3
C. 4 D. 5

19. Transcatheter arterial embolisation is a good option in the treatment of hepatocellular carcinoma in following situation:
A. Serum bilirubin > 3 mg%
B. Tumour thrombus in the main portal trunk
C. Early hepatocellular carcinoma
D. Inoperable tumours

20. Which of the following is true criteria for secondaries in the liver?
A. Single lobe enlargement
B. Rounded border
C. Soft in consistency
D. Nodular liver

21. Commonest gastrointestinal tract primary in a nodular liver secondaries is from:
A. Stomach
B. Pancreas
C. Gall bladder
D. Duodenum

22. Portal hypertension is defined as a portal venous pressure exceeding _____ mmHg.
A. 5 B. 10
C. 15 D. 20

23. Portal vein normally carries _____ % of blood supply to liver.
A. 25 B. 50
C. 75 D. 100

24. Causes of bulky secondaries include the following *except*:
A. Malignant melanoma
B. Carcinoid tumours
C. Colloid carcinoma rectum
D. Testicular tumours

25. Which of the following is true about the use of vasopressin in variceal bleeding?
A. 20 units is diluted in 200 ml of saline and given over 20 min
B. Causes coronary vasodilatation
C. Is a powerful inotrope
D. Is a splanchnic vasodilator and reduces pressure in the splanchnic vessels

26. Gastric transection of Tanner is an operation performed for:
A. Carcinoma stomach
B. Carcinoma pancreas
C. Portal hypertension
D. Hepatocellular carcinoma

ANSWERS

1 D	2 C	3 A	4 D	5 D	6 A	7 A	8 C	9 D	10 A
11 B	12 C	13 C	14 A	15 D	16 A	17 C	18 B	19 D	20 D
21 A	22 C	23 C	24 D	25 A	26 C				

25

Gall Bladder and Pancreas

- Surgical anatomy
- Physiology
- Congenital anomalies
- Gall stones disease
- Acute cholecystitis
- Chronic cholecystitis
- Obstructive jaundice
- Stricture CBD
- Sclerosing cholangitis
- Choledochal cyst
- Caroli's disease
- Chronic pancreatitis
- Cholangiocarcinoma
- Congenital biliary atresia
- Carcinoma of gall bladder
- Carcinoma of pancreas
- Endocrine tumours
- Acute pancreatitis
- Pseudocyst
- Annular pancreas
- Ectopic pancreas
- Cystic fibrosis
- Pancreatic divisum
- Pancreatic fistula
- White bile
- Pancreatic ascites
- What is new?/Recent advances

INTRODUCTION

- One may wonder why gall bladder and pancreas are discussed together!! Yes, biliary tract (gall bladder is an important part of that) ends after joining with pancreatic duct by opening into 2nd part of duodenum.
- Gall stones, when they block/pass through ampulla of Vater, produce acute pancreatitis.
- An important surgical condition—surgical jaundice or obstructive jaundice can be discussed better only if you know gall bladder and pancreas.
- Any surgery involving sphincter of Oddi in the form of sphincteroplasty/or removal (as in Whipple's pancreaticoduodenectomy), the gall bladder does not function and hence it needs to be removed.
- Embryologically, liver, biliary tree, ventral pancreas, gall bladder—all developed from a diverticulum on the ventral aspect of the foregut of the embryo.
- And gall bladder will be palpable in cases of periampullary/carcinoma head of the pancreas.

SURGICAL ANATOMY OF THE GALL BLADDER AND BILE DUCTS

Gall bladder is a pear or globular shaped organ present in the right hypochondrium on the inferior surface of the liver, situated in the gall bladder fossa. It is about 8–12 cm long.

- **Fundus:** It is the dilated portion of the gal lbladder adherent to undersurface of liver from which it can be separated easily.
- **Neck:** The narrow angulated distal portion of the neck is called ***Hartmann's pouch***—common site where stones occur and tend to stay for a long time (also called infundibulum of gall bladder) (Fig. 25.1).
- Gall bladder drains into the common bile duct (CBD) through ***cystic duct***, which is 3 cm long. It is lined by cuboidal epithelium. There are prominent mucosal folds within the cystic duct due to the presence of prominent

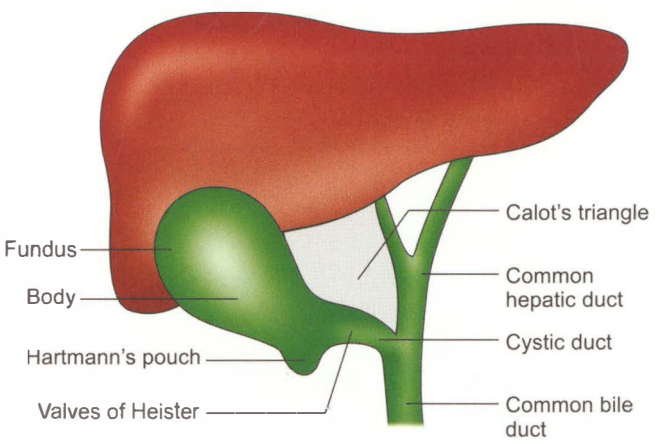


Fig. 25.1: Surgical anatomy of the gall bladder

circular muscle fibres underneath. Its lumen is usually 1–3 mm in diameter. Contraction of gall bladder produces a functional valve called ***valve of Heister*** which prevents the migration of stone into the CBD. The wall of cystic duct is surrounded by a sphincter structure called ***sphincter of Lutkens***. A spiral fold keeps cystic duct open for drainage of bile.

Cholecystohepatic triangle or Calot's triangle boundaries

- **Lateral:** Cystic duct and gall bladder
- **Medial:** Common hepatic duct
- **Above:** Inferior surface of right lobe of the liver.

It is an important landmark in the identification of cystic duct, and cystic artery during cholecystostomy so as to avoid damage to extrahepatic biliary tree.

Contents

- Right hepatic artery and its branch, the cystic artery
- Cystic lymph node of Lund.

Blood supply of gall bladder

Cystic artery, a branch of right hepatic artery arises behind the common bile duct. Soon, it branches out over the surface of gall bladder. ***Cystic artery is an end artery*** (Fig. 25.2). Multiple small veins from the surface of gall bladder join the liver surface. There is also a cystic vein, from the neck of gall bladder draining into portal vein directly. This explains early spread of gall bladder malignancy to the liver.

Lymphatics

1. Subserosal and submucosal lymph nodes drain into ***cystic lymph node of Lund*** and from here they drain into nodes in the hilum of liver and coeliac nodes.
2. **Subserosal lymphatic vessels** of gall bladder are also connected to **subcapsular lymph channels** of liver, which accounts for frequent spread of carcinoma gall bladder to the liver.

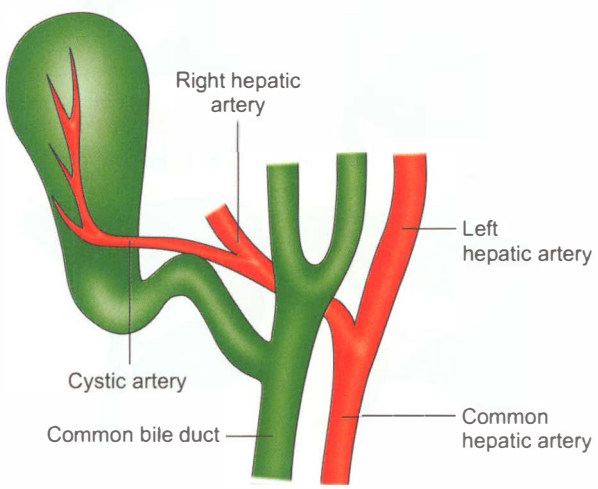


Fig. 25.2: Blood supply of the gall bladder

Anatomy of the bile ducts

- **Common hepatic duct (CHD)** is formed by the union of right and left hepatic ducts. It is 3 cm long, receives cystic duct and continues as common bile duct (CBD).
- **Common bile duct** is about 8 cm long. It has four parts: Supraduodenal, retroduodenal, infraduodenal and intraduodenal. Along with pancreatic duct, it forms ampulla of Vater. Controlled by sphincter of Oddi, it ends by an opening into the second part of duodenum (Fig. 25.3).

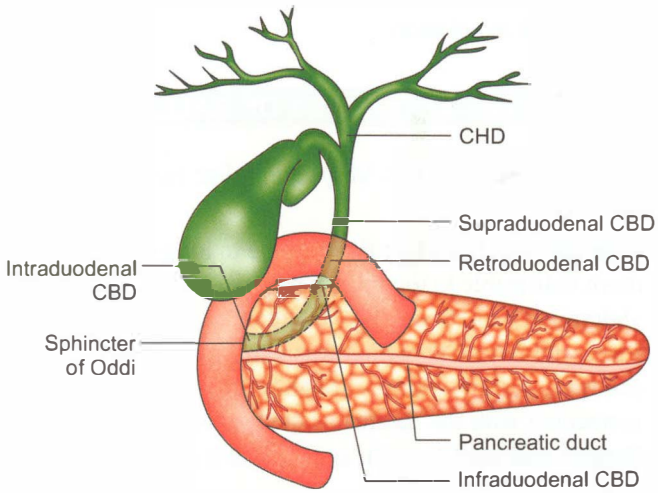


Fig. 25.3: Surgical anatomy of the bile ducts. Note how close gall bladder is to the duodenum (chances of injury in laparoscopic cholecystectomy)

PHYSIOLOGY

Functions of the gall bladder

- **Reservoir for bile:** Bile excreted by the liver is stored in the gall bladder as total of about 500 to 1000 ml per day. At fasting, the tone of sphincter of Oddi is high. Food contents in the duodenum stimulates release of cholecystokinin, which causes gall bladder to contract.
- **Concentration:** Bile is 98% water. Due to active absorption of water, sodium chloride and bicarbonate, bile gets concentrated 5–10 times. Thus, a relative increase in bile salts, bile pigments, cholesterol and calcium occurs.
- **Mucus secretion:** It secretes about 20 ml/mucus per day. Obstruction to the cystic duct causes mucocoele of the gall bladder.

Bile

- Secreted from hepatocytes
- pH is more than 7.0

- 500–1000 ml/day, 98% is water
- Concentrated in gall bladder because of absorption of water. Capacity of gall bladder is 40–50 ml.
- Fatty food stimulation releases cholecystokinin, which stimulates gall bladder to contract and at the same time, sphincter of Oddi to relax.
- It also has inorganic ions (more than plasma) and hence, severe electrolyte imbalance is seen in biliary fistula.
- Cholesterol, synthesised in the liver, gives rise to bile acids—cholic and chenodeoxycholic acids. They are metabolised in the colon to deoxycholic acid and lithocholic acids.
- Main function of bile acids in the bile is to maintain cholesterol in solution.

CONGENITAL ANOMALIES OF GALL BLADDER

Absence of gall bladder: Very rare; other variations include (Fig. 25.4):

A. **Floating gall bladder:** Results due to long mesentery. It is more vulnerable to torsion—a rare cause of recurrent upper abdominal pain. Such a gall bladder can be easily removed.

B. **Phrygian cap:** Cap which was worn by people of Phrygia (ancient Asian country, Mongolia). It is an anomaly connected with the fundus of the gall bladder.

C. **Double gall bladder:** The second one is always intrahepatic (rare).

D. **Absence of cystic duct:** Cholecystectomy becomes difficult. There are high chances of injury to the common bile duct.

E. **Low insertion of cystic duct:** Cystic duct opens into the common bile duct near the ampulla. This anomaly should be kept in mind when operating on cases of obstructiv jaundice.

F. **An accessory or aberrant cholecystohepatic duct** i present in about 10% of the patients. It may be the cause o significant bile leakage after cholecystectomy. It is th segmental duct that joins biliary system outside the live instead of within it.

G. Diverticulum of gall bladder

H. Cystic duct joining right hepatic duct

I. **Anomalies of blood supply**

1. Very, very tortuous hepatic artery: Caterpillar turn o Moynihan's hump. It runs in front of the origin of cysti duct.
2. Cystic artery is given anteriorly from right hepati artery.

GALL STONE DISEASE (CHOLELITHIASIS)

Aetiology

1. Metabolic causes

- Cholesterol is produced from the liver which gives rise tc bile acids. Cholesterol is insoluble and it must be transportec within the bile salt micelles and phospholipid (lecithin) vesicles. Normal ratio of bile acids: cholesterol is 25:1 (Fig. 25.5).
- This ratio is necessary to maintain the cholesterol in liquic form by forming micelles. When the ratio drops down tc 13:1 (which is called critical ratio), cholesterol crystals wil nucleate and stones will form.
- Obesity, high calorie diet and medications which increase cholesterol secretion can result in stone formation.

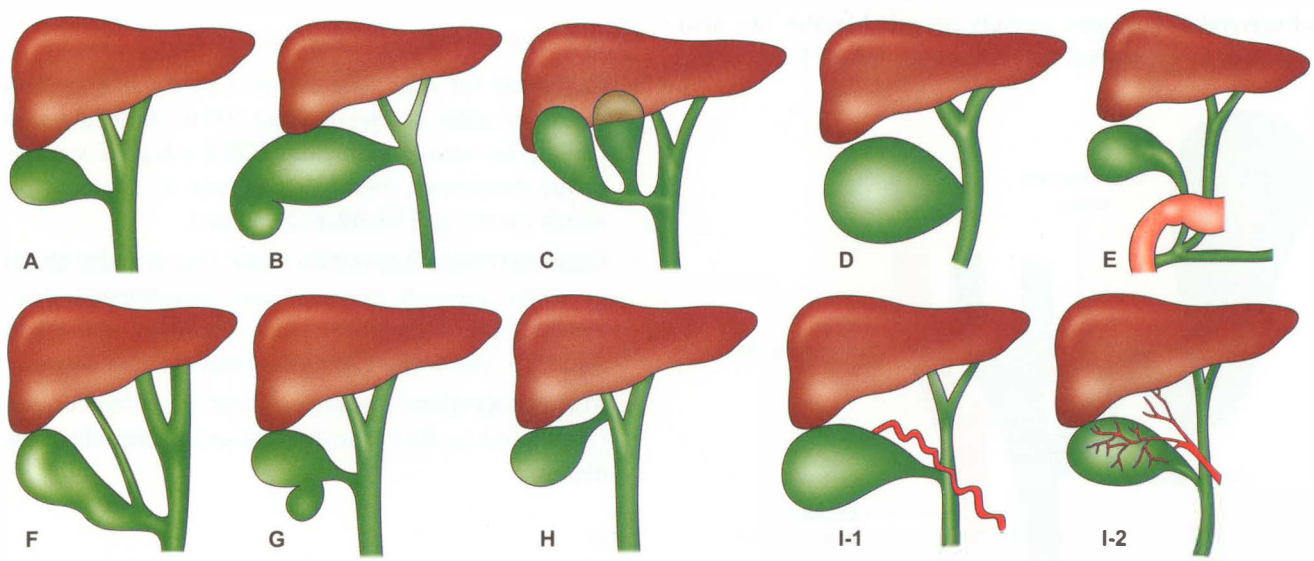


Fig. 25.4: Congenital anomalies of the gall bladder (*see* text A to I)

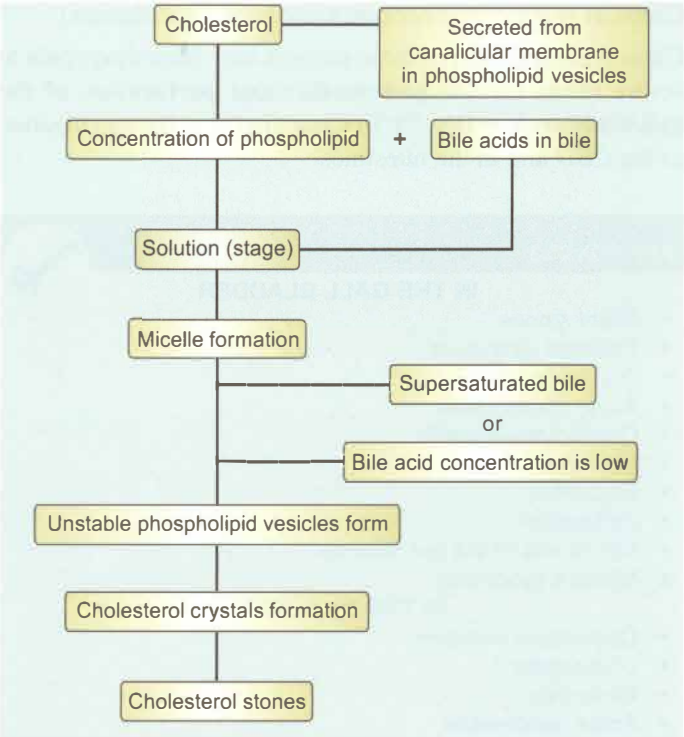


Fig. 25.5: Flow chart showing formation of cholesterol stones

2. Infection

- It is the most common cause responsible for a gall stone in 80% of patients. Sources of infection are tonsils, tooth, bowel, etc.
- Organisms such as *E. coli,* Proteus, anaerobic organisms, Streptococci, etc. reach the gall bladder wall through the blood stream and form a focus/nidus around which cholesterol and bile salts get precipitated.
- Over a period of many years, this results in a mixed stone. They are usually multiple and occur in infected bile.

PEARLS OF WISDOM

"A gall stone is a tomb stone erected in the memory of organisms within it."—Lord Moynihan

3. Bile stasis and decreased bile acid pool

- Pregnancy, oestrogens, following vagotomy and prolonged total parenteral nutrition are associated with bile stasis.
- They are prone to mixed stones as a result of bile stasis (Key Box 25.1).

KEY BOX 25.1

CAUSES OF DECREASED BILE ACID POOL

1. Cirrhosis of liver—pigment stones
2. Gastrectomy
3. Ileal resection
4. Malabsorption
5. Obesity
6. Hypercholesterolaemia

4. Haemolytic anaemia

- Examples: Hereditary spherocytosis, sickle cell anaemia.
- Bilirubin production is increased because of **increased break down of RBCs.** Since the production is more, they cannot conjugate with glucuronic acid, which is produced at normal levels.
- Such **unconjugated bilirubin** combines with calcium and is excreted in the biliary tree resulting in calcium bilirubinate stones (pigment stones) not only in the gall bladder but also in the entire ductal system.

5. Saint's triad

Gall stones (can occur along with two other conditions mentioned below)

- Diverticulosis of colon
- Hiatus hernia

6. Parasitic infestation

- In Oriental countries, *Clonorchis sinensis* (Chinese liver fluke) infestations can cause stone in the biliary tree.
- *Ascaris lumbricoides* in the biliary tree may produce stones in our country (India).

7. Due to abnormal mucus

It is produced in **congenital cystic fibrosis.** Gall stones occur in these children due to impairment of bile flow.

Other diseases associated with gall stone disease

- Diabetes mellitus
- Type IV hyperlipoproteinaemia
- Cirrhosis of liver
- Fistulae on treatment with total parenteral nutrition
- Gastric surgery

Risk factors for gall stone disease

(Key Box 25.2 and Fig. 25.6)

KEY BOX 25.2

RISK FACTORS

- Female sex
- Obesity
- Maturity onset diabetes
- Age > 40 years

Types of gall stones (Figs 25.7 to 25.16)

1. Cholesterol stones

- **Constitutes about 10% of the gall stones.**
- Occur in patients with increased cholesterol levels.
- Fatty women are commonly affected.
- It is single, solitary, occurs in aseptic bile. Sometimes they can be multiple. Precipitation of cholesterol gives rise to stone.

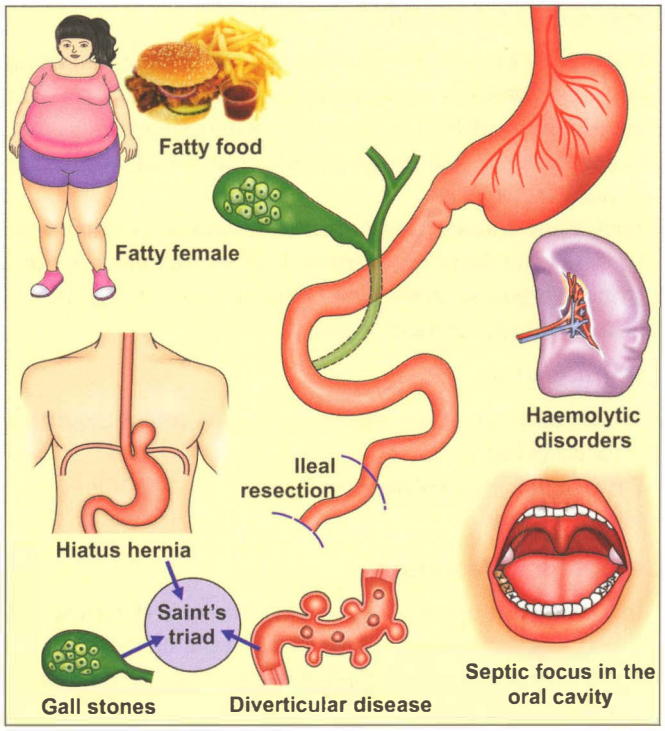


Fig. 25.6: A few risk factors for gall stone disease

- Such stones can be silent for many years. They are radiolucent.
- Pigment can also get precipitated along with cholesterol.

2. Brown pigment stones

- **Rare in gall bladder, occurs in bile duct.**
- Composed of calcium bilirubinate, calcium palmitate and calcium stearate + cholesterol.
- Occur due to bile stasis caused by foreign bodies, endoprosthesis, *Clonorchis sinensis* and *Ascaris lumbricoides*.

3. Mixed stones

- They constitute about 80% of gall stones.
- They contain alternating layers of cholesterol and pigment with epithelial debris or vegetations, from infective organisms.
- They are multiple, small, faceted by mutual pressure.

4. Pigment stones

- They are found in 5 to 10% of patients.
- They are calcium bilirubinate stones.
- Commonly occur due to haemolysis. Hence, they are black, multiple, small, irregular concretions or sludge particles.
- For reasons not clear, **cirrhotic patients have increased incidence of black pigment stones.**
- Bacteria also have a major role to play in the formation of pigment stones. Patients with pigment stones have more sepsis than patients with cholesterol stones.

Clinical features (complications of gall stones)

Clinical presentation of these patients vary from **dyspepsia** to severe forms such as **pancreatitis and perforation of the gall bladder** (Key Box 25.3). Classified as in the gall bladder in the CBD and in the intestines.

KEY BOX 25.3

IN THE GALL BLADDER

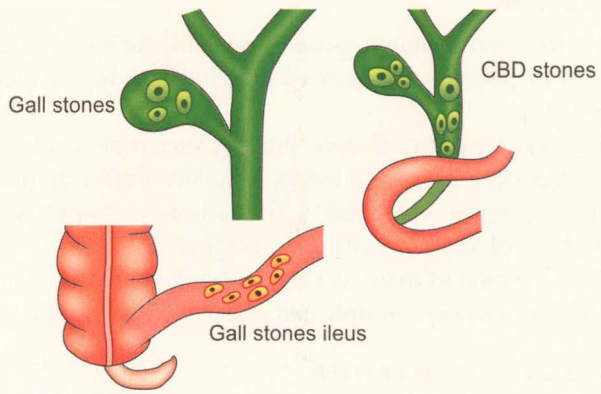
- Silent stones
- Flatulent dyspepsia
- Gall stone colic
- Acute cholecystitis
- Chronic cholecystitis
- Mucocoele
- Empyema
- Perforation
- Carcinoma of the gall bladder
- Mirizzi's syndrome

IN THE BILE DUCT

- Obstructive jaundice
- Cholangitis
- White bile
- Acute pancreatitis

IN THE INTESTINE

Acute intestinal obstruction (gall stone ileus)



COMPLICATIONS IN THE GALL BLADDER

Silent stones

- This is usually a single, silent, cholesterol stone which is symptomless.
- It is accidentally discovered, may be by an ultrasound or plain X-ray abdomen (since calcium content is low in a cholesterol stone, it is very rarely visible in a plain X-ray).
- This stone rarely causes obstructive jaundice.
- Hence, it is left alone without treatment.

Flatulent dyspepsia

If an obese woman (fatty, fertile, flatulent, female in forties) complains of gaseous distention, intolerance to fatty food and discomfort in the abdomen, heartburn and belching, she probably has gall stones. These patients benefit from cholecystectomy.

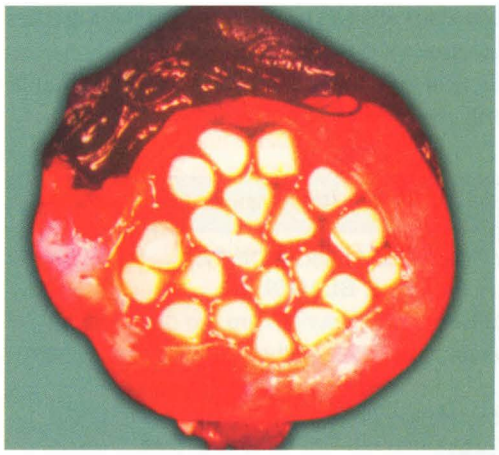


Fig. 25.7: Mixed stones—cause of flatulent dyspepsia

Fig. 25.8: Pigment stones—case of hereditary spherocytosis

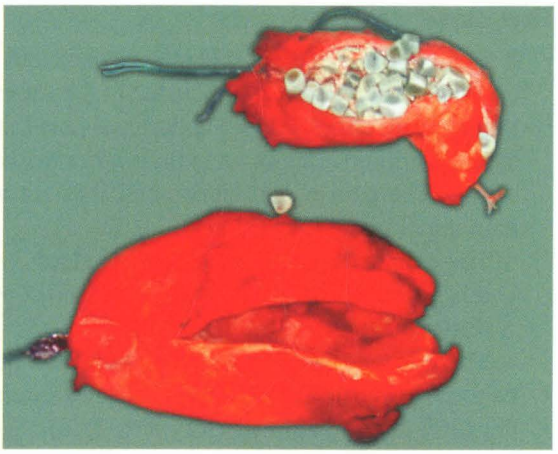
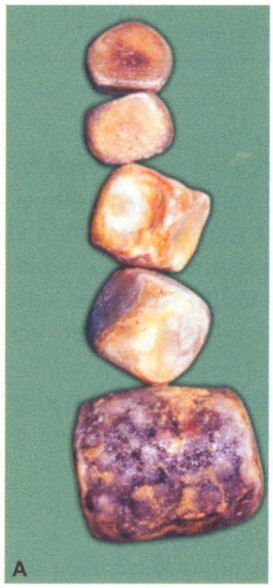


Fig. 25.9: Double GB with stones



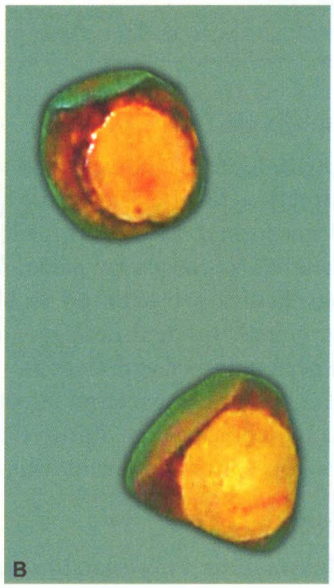


Fig. 25.10: Mixed stones—observe faceting—cause of gall stone colic

Fig. 25.11: Gall stones—see the colour

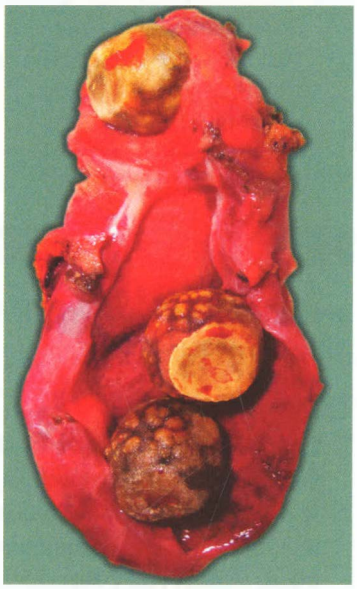


Fig. 25.12: Contracted thick-walled gall bladder

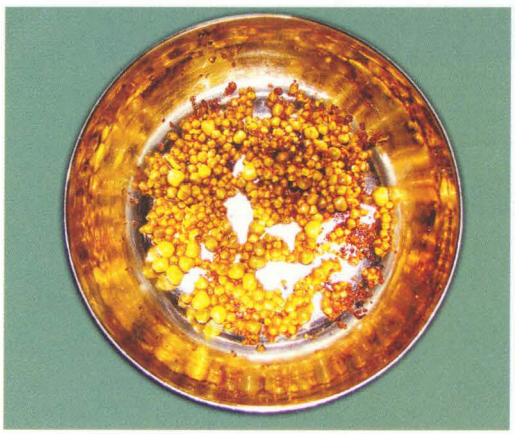
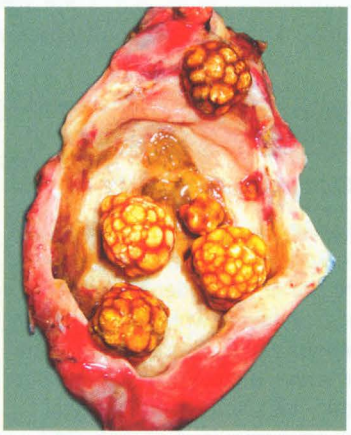
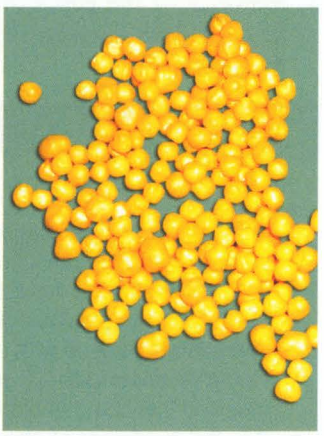


Fig. 25.13: Multiple 2 mm stones. This lady had 3 attacks of gall stone pancreatitis

25.14: Gallbladder was opened to extract stones





Figs 25.15 and 25.16: Gall stones: Multiple stones are responsible for obstructive jaundice

Gall stone colic

- It usually occurs at night wherein a stone tends to block the cystic duct or neck of gall bladder in the supine position.
- It is a severe colicky upper abdominal pain felt in the right hypochondrium, may shoot to the back or between shoulder blades. The pain is continuous and lasts for a few hours. Pain may radiate to chest also.
  - The pain is due to spasm of gall bladder
  - It is associated with vomiting due to reflex pylorospasm, restlessness and sweating.
  - There is tenderness in the right hypochondrium.
- Pain may last for a few minutes to a few hours.
- For differential diagnosis (Key Box 25.4).

KEY BOX 25.4

DIFFERENTIAL DIAGNOSIS OF GALL STONE COLIC

- Chronic duodenal ulcer
- Reflux oesophagitis (some cause can present as precordial chest pain)
- Pancreatitis
- Myocardial infarction

ACUTE CHOLECYSTITIS

Definition: Acute bacterial inflammation of the gall bladder with or without stone.

Types

1. **Calculous:** Obstructive cholecystitis. It is the commonest variety. Calculi cause bile stasis.
2. **Acalculous:** Nonobstructive cholecystitis. It is not uncommon and is seen in patients who are recovering from major illness (Key Box 25.5).
3. Acute emphysematous cholecystitis.

Bacteriology of acute cholecystitis

1. Majority of the cases of calculous cholecystitis are due to organisms such as *E. coli, Streptococci, Salmonella, Klebsiella,* etc.

KEY BOX 25.5

PERCUTANEOUS CHOLECYSTOSTOMY

- In life-threatening situations with severe sepsis due to gall stones, percutaneous cholecystostomy seems to be a very good alternative to save the life of the patient.
- Indicated in acalculous cholecystitis in which the patient's condition is serious with sepsis with co-morbid conditions.
- Using ultrasound or CT guidance, a pigtail catheter can be inserted into the gall bladder, ideally transperitoneally.
- It is the treatment of choice for acalculous cholecystitis.
- 3–4 days later, when sepsis improves, laparoscopic/open cholecystectomy can be done.

2. Typhoid fever can also cause **Typhoid Cholecystitis** around 2nd week of infection (Key Box 25.6).
3. *Clostridial* infection of the gall bladder produces acute cholecystitis with toxaemia.

KEY BOX 25.6

TYPHOID INFECTION OF THE GALL BLADDER

- *Salmonella typhi* or *S. typhimurium* are the organisms
- Acute cholecystitis can occur in the 2nd week of typhoid fever.
- Long lasting infections—chronic cholecystitis can occur
- Bacilli can be present in the bile for a long time
- Obstruction by a pre-existing stone or any other cause
- Necrosis of GB, ulceration, perforation can be dangerous
- Local tenderness, on the right side of abdomen, guarding can occur.
- Antibiotics against Salmonella must be given

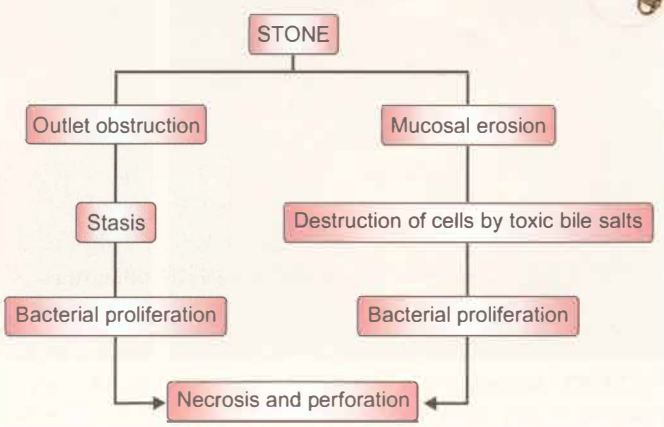
Pathogenesis (Key Box 25.7)

- Acute calculous cholecystitis appears to be caused by obstruction to bile flow from gall bladder by stone or oedema formed as a result of local mucosal erosion and inflammation caused by stone. **Once mucosa is eroded, tissue planes are exposed to bile salts.** Toxic bile salts destroy cells by their detergent action leading to necrosis and perforation of gall bladder.
- At the same time, bacterial infection adds to the morbidity of acute cholecystitis. Positive bile cultures are found in 70% of cases of acute calculous cholecystitis.

Pathology

1. **Inflammation:** Entire gall bladder is inflamed, swollen and is friable. When the inflammatory exudate surrounding the gall bladder collects under the diaphragm, it results in pain radiating to the right shoulder (C3, 4) due to phrenic nerve

KEY BOX 25.7



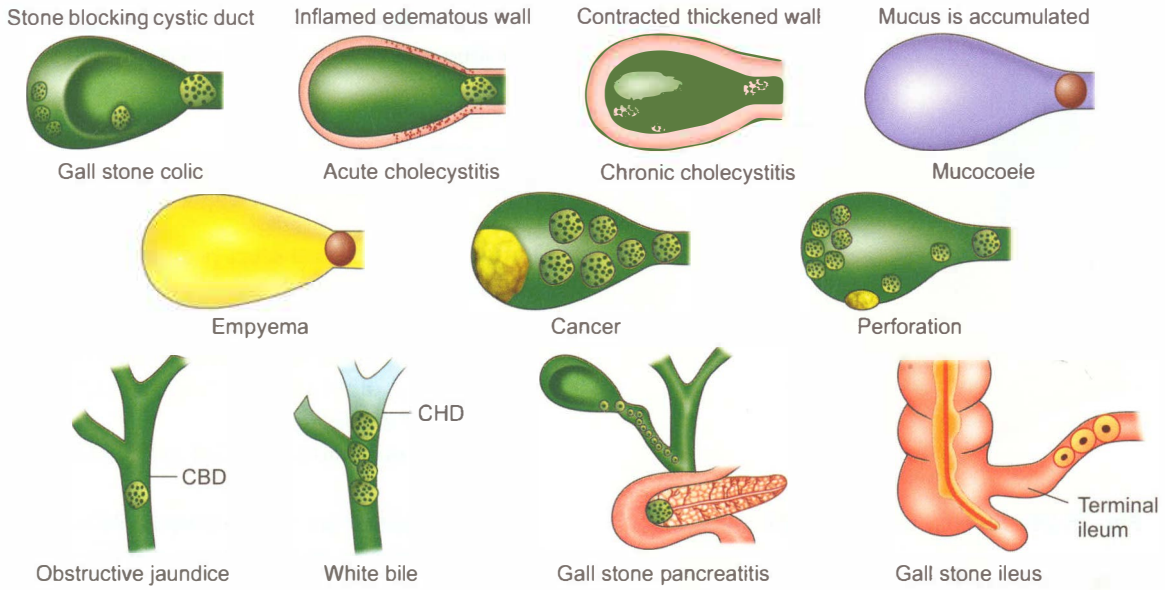


Fig. 25.17: Diagrammatic representation of the complications of gall stones. They are discussed in the subsequent pages

irritation. It may undergo complete resolution with antibiotic therapy but such recurrent attacks are common at a later date (Fig. 25.17).

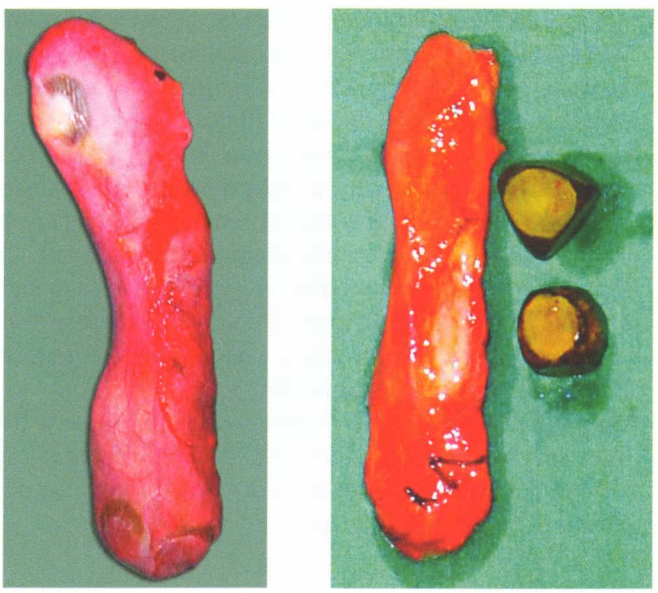
2. **Perforation:** Extensive ulcerations of gall bladder may result in perforation with biliary peritonitis and carries a very high mortality rate. **Perforation** can occur when the stone is impacted in the **Hartmann's pouch**.
3. **Obstruction** to the neck of gall bladder results in **mucocoele** or **pyocoele** (empyema). Empyema of the gall bladder can occur in diabetic patients and is associated with high grade fever, chills, rigors and even septicaemia.
4. **Gangrene** of gall bladder can occur if the blood vessels get thrombosed. All these features are more in an obstructive variety.
   - If there is clostridial infection as can occur in diabetics because of extensive gas production in biliary tree and associated toxicity, perforation is likely even without a stone.

Clinical features

- A fatty, fertile, female is the typical victim who presents with severe upper abdominal pain (Figs 25.18 and 25.19). The pain is colicky in nature and more prolonged because of inflammation. Severe nausea and vomiting are present. In the initial phase, there is low grade fever, except in clostridial infection where there is high grade fever.
- You may be surprised to find a young boy, a girl or even a child with gall stones—suspect haemolytic anaemias.

Signs

1. ***Murphy's sign***
   - Keep the fingers in the right hypochondrium and ask the patient to take a deep inspiration.
   - At the height of inspiration there is a sudden catch in the inspiration.
   - It is due to inflamed gall bladder coming in contact with the abdominal wall under the fingers and producing pain. This is called **Murphy's sign positive**. It is a diagnostic sign of acute cholecystitis (Fig. 25.20).
2. ***Boas' sign:*** An area of hyperaesthesia between 9th and 11th ribs posteriorly on the right side is a feature.
3. Upper abdominal **guarding, rigidity**.
4. **Vague mass** consisting of inflamed gall bladder, omentum, inflammatory exudate can be felt at times. Hence, even if a perforation occurs, generalised peritonitis is uncommon.



Figs 25.18 and 25.19: Large 2 stones blocking lumen—cause for severe colic

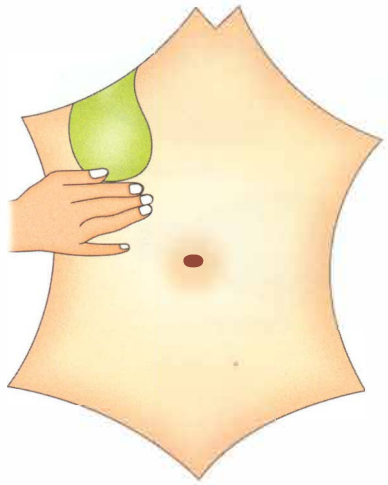


Fig. 25.20: Eliciting Murphy's sign

Differential diagnosis (DD) (Fig. 25.21)

1. **Perforated peptic ulcer:** Severe sudden pain, severe tenderness in the right hypochondrium, guarding and rigidity caused by perforated peptic ulcer, can mimic acute cholecystitis. Obliteration of liver dullness, coffee ground vomitus, generalised guarding and rigidity clinches the diagnosis of perforated duodenal ulcer.
2. **Acute pancreatitis:** A severe pain in upper abdomen, tenderness in the right hypochondrium and epigastrium mimic cholecystitis. One should remember that pain of pancreatitis is more severe and classically radiates to back.
3. **High retrocaecal appendicitis:** Especially when appendix is in the subhepatic position. Once inflammatory fluid spreads in the general peritoneal cavity, there will be more difficulty in diagnosing clinically.
4. **Amoebic liver abscess:** Can also mimic very closely. It is more common in male alcoholics. Liver is enlarged and one can feel the round lower border of liver very closely. Liver will be extremely tender.
5. **Lobar pneumonia (basal):** Can cause right hypochondriac guarding and rigidity. It is a referred pain.

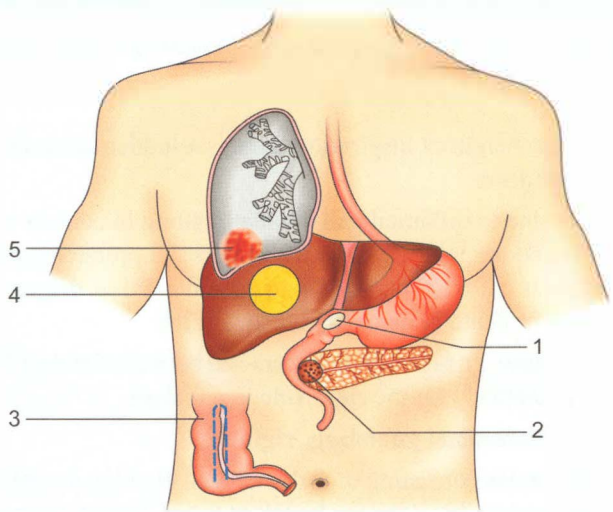


Fig. 25.21: Differential diagnosis of acute cholecystitis (*see* text)

Investigations

1. **Total WBC** count is always raised.
2. **Blood and urine sugar** estimation to rule out diabetes mellitus.
3. **Plain X-ray** abdomen erect position (Fig. 25.22)
   - Gall stones can be demonstrated in 10% of the patients as radio-opaque shadows in the right hypochondrium. In lateral view, the stone is seen in front of vertebral bodies.
   - To rule out other causes such as perforated peptic ulcer (air under diaphragm).
   - Rarely, it may show calcified gall bladder (porcelain gall bladder).

   10% Gall stones are radio-opaque, 90% are radiolucent. Centre of stones may contain radiolucent gas, either triradiate (**Mercedes Benz sign**) or biradiate **(Sea Gull sign).**
4. **Emergency ultrasonography** (Fig. 25.23)
   - To demonstrate stones, which cast **posterior acoustic shadow**.
   - Success rate is > 95%
   - It can demonstrate inflamed, thickened organ, in cases of acalculous cholecystitis.
   - Demonstration of Murphy's sign, with the help of ultrasonography is possible which adds to the diagnosis.
   - Ultrasound can also measure gall bladder function by using ultrasonic dimensions of the gall bladder.
   - It can detect gall bladder polyps.
5. **HIDA scan/PIPIDA scan** (Fig. 25.24)
   - HIDA is **hepatic iminodiacetic acid**.
   - ^{99m}Tc labelled **HIDA** agent is excreted in the biliary tree, within one hour following IV administration.

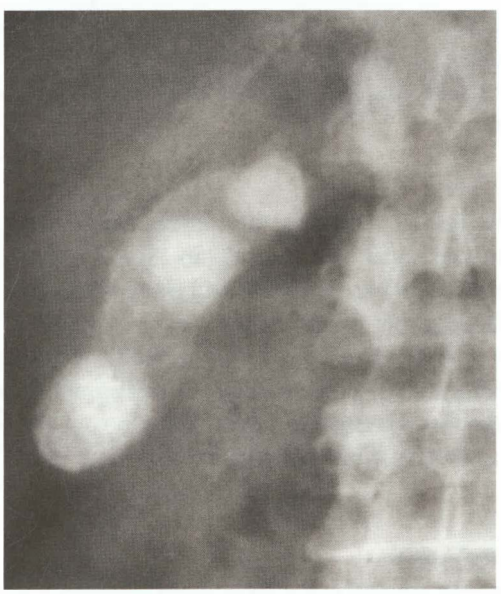


Fig. 25.22: Plain X-ray abdomen showing radio-opaque shadows

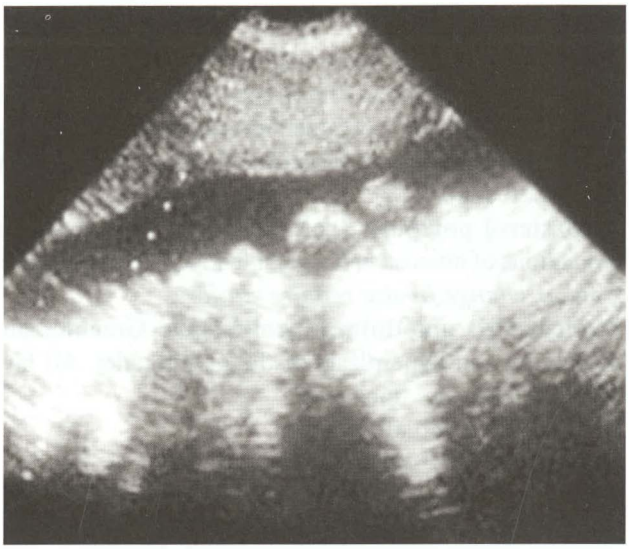


Fig. 25.23: Ultrasonography showing posterior acoustic shadows

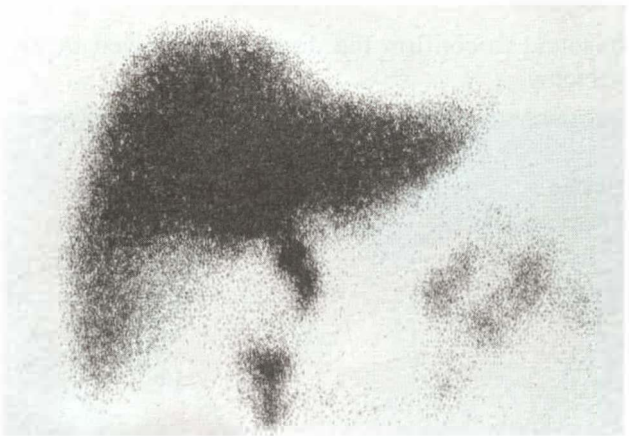


Fig. 25.24: HIDA scan showing nonvisualisation of gall bladder—a case of acute cholecystitis (CT is more specific)

- In acute cholecystitis, even though the dye is excreted in the biliary tree, it does not enter the gall bladder due to oedema of the cystic duct. Hence, nonvisualisation of gall bladder is diagnostic of acute cholecystitis.
- Its importance lies in the **diagnosis of acalculous cholecystitis** and when diagnosis is in doubt.

6. **CT Scan:**
   - It is done when ultrasound findings are not clear
   - It not only diagnoses gall stones, but also detects other complications such as perforation, stones in the CBD, etc.
   - Renal halo sign due to fluid around
   - Obliteration of psoas shadow
   - Air fluid level in duodenum are the features.

Treatment: School of thought

I. Conservative treatment is followed in majority of the cases (60 to 70%)

1. **Admission** in the hospital
2. **Aspiration** with Ryle's tube: Aspiration of HCl decreases the stimulus to the secretion of bile. Spasm of gall bladder may come down.
3. **Antispasmodics:** Injection morphine 8–10 mg IM as analgesic along with injection atropine 0.6 mg to relieve spasm of sphincter of Oddi.
4. **Antibiotics:** Broad spectrum antibiotics are given against gram +ve, gram –ve and anaerobic organisms. Cefazolin, cefuroxime or amikacin are the drugs of choice. The patient is kept nil orally for 2–3 days and during this time IV fluids are given.

After 2–3 days, pain comes down, signs (tenderness) disappear and abdomen becomes soft. Ryle's tube is removed, clear oral fluid is given for 2–3 days followed by soft diet. After 6 weeks, the patient is advised to undergo elective cholecystectomy. Reason for conservative treatment is in majority of cases, inflammation will settle down.

II. Early cholecystectomy

- Patients in the first group need two admissions, cost is increased and return to work is also delayed.
- Hence if a surgeon is experienced and the set up is good, one can proceed to early cholecystectomy from 2nd day to 7th day.
- It has been proved that even though gall bladder is inflamed, complications are no way more than elective cholecystectomy in the hands of experienced surgeon while performing laparoscopic cholecystectomy.
- Thus, if a firm preoperative diagnosis is established and some of the comorbid conditions are corrected (diabetes, hypertension, etc.), surgery can be done safely. This is called early cholecystectomy.

III. Emergency cholecystostomy

- About 10% of cases of acute cholecystitis require emergency cholecystostomy.
- In these patients, high grade fever, sepsis, shock, high leukocyte count are the deciding factors.
- Acalculous cholecystitis and perforated gall bladder with peritonitis are definitely strong indications for emergency cholecystectomy.

IV. What is prophylactic cholecystectomy?

It means removal of gall bladder with stones without symptoms (Key Box 25.8).

Prognosis

- Overall death rate is 3–5%.
- Contributing factors for death are diabetes, age above 60 years, cardiovascular or pulmonary disease.
- Uncontrolled sepsis, intra-abdominal abscess.

KEY BOX 25.8

PROPHYLACTIC CHOLECYSTECTOMY

- Diabetic patients
- Congenital haemolytic anaemia
- Patients undergoing bariatric surgery

CHRONIC CHOLECYSTITIS

- Recurrent attacks of cholecystitis will convert the gall bladder into a fibrosed, nonfunctioning, contracted, shrunken, small gall bladder. Gall bladder wall is grossly thickened. Stones are invariably present. Such patients present with classical fatty food intolerance. Murphy's sign is positive.
- They are diagnosed by ultrasound which reveals a small, contracted gall bladder. Otherwise, oral cholecystography (OCG) can be done to know the function of the gall bladder.

Treatment

Cholecystectomy (Fig. 25.25)

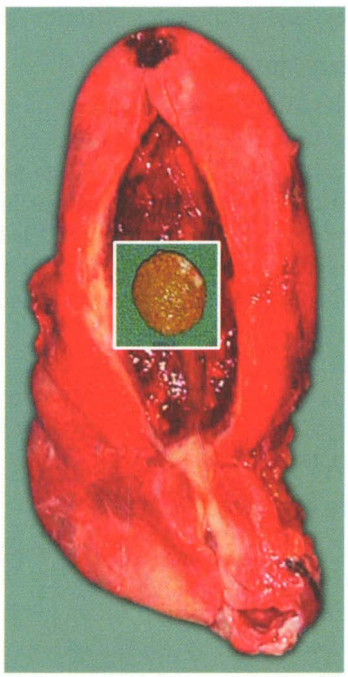


Fig. 25.25: Cholecystectomy specimen—observe thickened gall bladder wall

CHOLECYSTOSES

Definition

Uncommon group of conditions affecting gall bladder in which there are chronic inflammatory changes and hyperplasia of all tissue elements.

KEY BOX 25.9

XANTHOGRANULOMATOUS CHOLECYSTITIS

- It is a pathological diagnosis
- Gall bladder is chronically thickened, irregular with extension of yellow xanthogranulomatous inflammation to adjacent organs.
- It is thought to be due to bile penetrating gall bladder wall

Types

1. **Cholesterosis** (Strawberry gall bladder): Aggregations o cholesterol crystals in the mucosa/submucosa. Yellov specks of cholesterol crystals are seen when gall bladde is opened (xanthogranulomatous) (Key Box 25.9 anc Figs 25.26 and 25.27).
2. **Cholesterol polyposis** (gall bladder polyp): Polypoida projections of mucosa in the gall bladder. ***Polyp longer thar 1 cm or change in size require surgery.***
3. **Cholecystitis glandularis proliferans:** Granulomatous thickening and hyperplasia of the gall bladder. All layers of gall bladder are thickened.
4. **Diverticulosis** of gall bladder
5. Gall bladder wall with ***fistula***

Clinical features

Dyspepsia, upper abdominal discomfort, Murphy's sign is positive.

Management

Ultrasound to confirm the diagnosis followed by cholecystectomy.

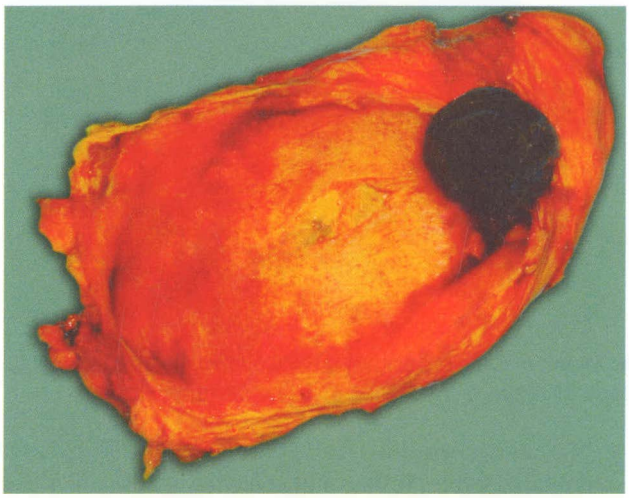


Fig. 25.26: Xanthogranulomatous cholecystitis with stone

Fig. 25.27: Xanthogranulomatous cholecystitis at surgery

MUCOCOELE (Figs 25.28 to 25.30)

- It occurs when there is a stone blocking the cystic duct and the bile is not infected.
- As a result of obstruction, all the bile within the gall bladder is absorbed and is replaced by the mucus secreted from gall bladder epithelium.
- Clinically, it results in a soft, fluctuant, globular mass in the right hypochondrium which moves with respiration. It needs cholecystectomy.

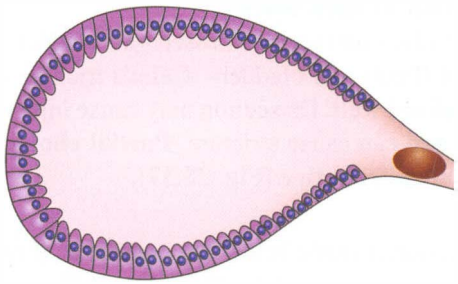


Fig. 25.28: Obstruction by stone

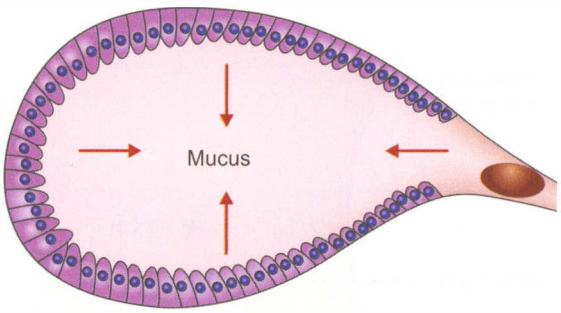


Fig. 25.29: Mucocoele

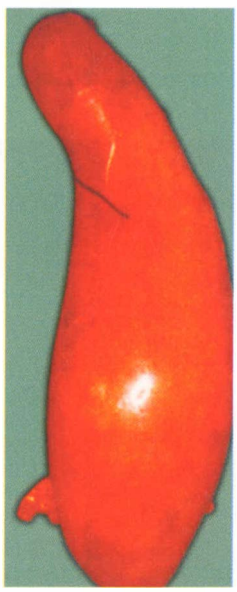


Fig. 25.30: Mucocoele

EMPYEMA AND PERFORATION OF GALL BLADDER

- These are uncommon. Impacted stone, diabetes, virulent organisms precipitate pyocoele and perforation.
- Patients present with high grade fever with chills and rigors, toxicity, high leukocyte count.
- Perforation can cause local abscess, if there are adhesions due to previous inflammation.
- Perforation into the general peritoneal cavity is rare but produces diffuse biliary peritonitis which has a high mortality rate.
- Urgent laparotomy, aggressive resuscitation, good antibiotic cover may help in reducing mortality. At laparotomy, removal of gall bladder is difficult. Hence, drainage of the pus and cholecystostomy with removal of gall stones can be done (Figs 25.31A and B).

3 Types (Fig. 25.32)

1. Localised perforation with pericholecystic abscess-managed by catheter drainage.

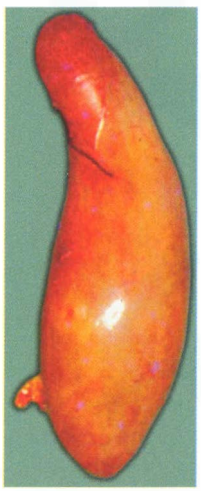


Fig. 25.31A: Empyema of gall bladder

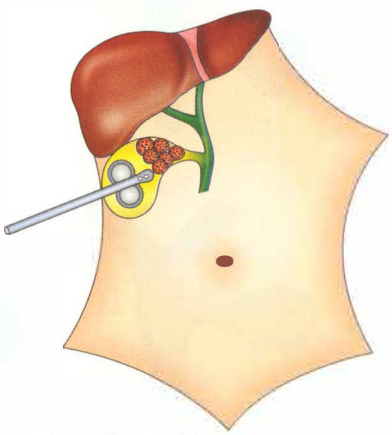


Fig. 25.31B: Percutaneous cholecystostomy for empyema of gall bladder

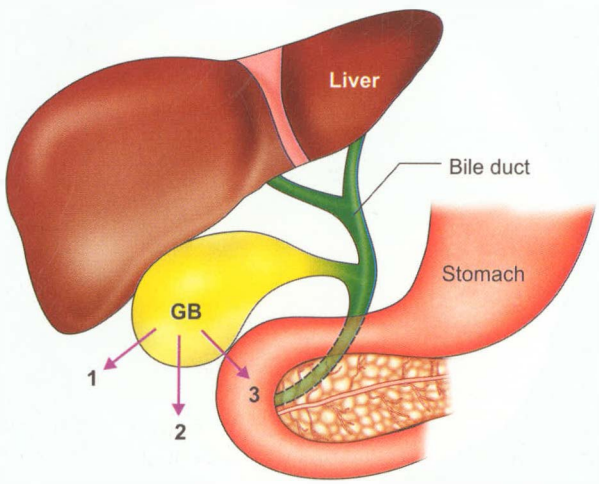


Fig. 25.32: Empyema of gall bladder—complications (*see* the text above)

2. Free perforation into peritoneal cavity and peritonitis—urgent laparotomy drainage.
3. Hollow viscous perforation (duodenum).

CARCINOMA OF GALL BLADDER (page 589)

Long-standing gall stones can bring about squamous metaplasia of gall bladder epithelium and can cause carcinoma of gall bladder. However, the incidence is very low. Hence, routine cholecystectomy is not advised for silent gall stones.

MIRIZZI SYNDROME (Figs 25.33 and 25.34)

Type I: Compression of CBD without lumen narrowing.
Type II: Compression of CBD with lumen narrowing.
Type III: Compression causing CBD wall necrosis.
Type IV: Stone ulcerating into CBD resulting in cholecysto-choledochal fistula.

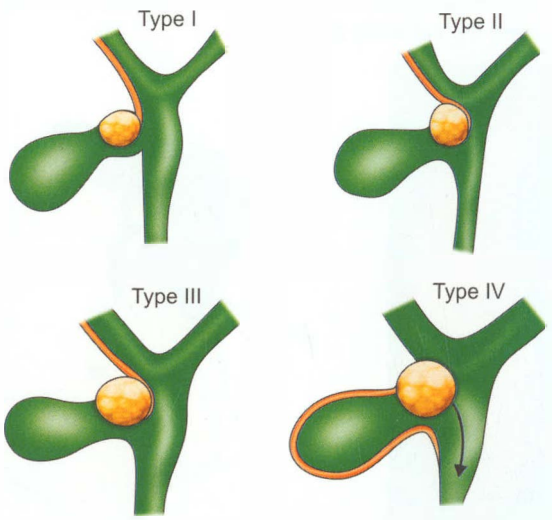


Fig. 25.33: Mirizzi syndrome

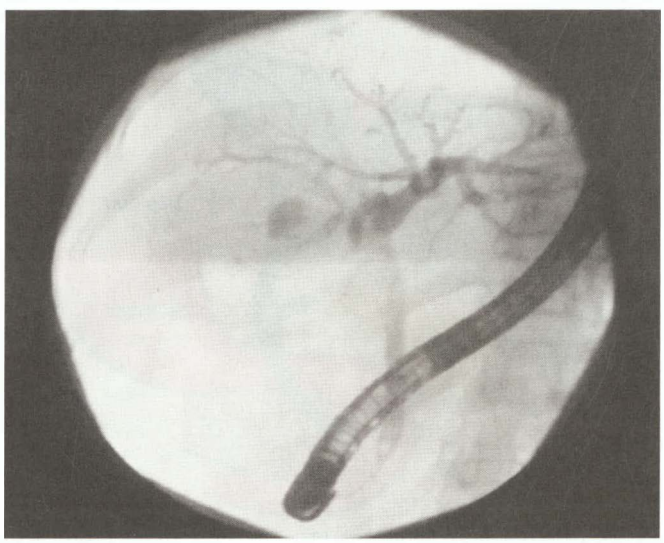


Fig. 25.34: ERCP showing Mirizzi syndrome

TREATMENT OF GALL STONES

ERCP (Fig. 25.34) followed by **open cholecystectomy** was the most popular method till recently, now replaced by laparoscopic cholecystectomy.

Laparoscopic cholecystectomy

It has become the most popular choice today. More than 95% of gall bladders can be removed through a laparoscope. Some principles and procedure of laparoscopic cholecystectomy are discussed below (Figs 25.35 and 25.36).

Conversion to open when:

- Very badly contracted, fibrosed gall bladder.
- Very difficult gall bladder—Calot's triangle anatomy is not defined well. Dissection may cause injury to the bile ducts and can cause stricture. **Partial cholecystectomy is a safe alternative** (Fig. 25.37).

Procedure

- 1 cm incision is made below the umbilicus, through which a pneumoperitoneum is maintained by CO_2 insufflation.

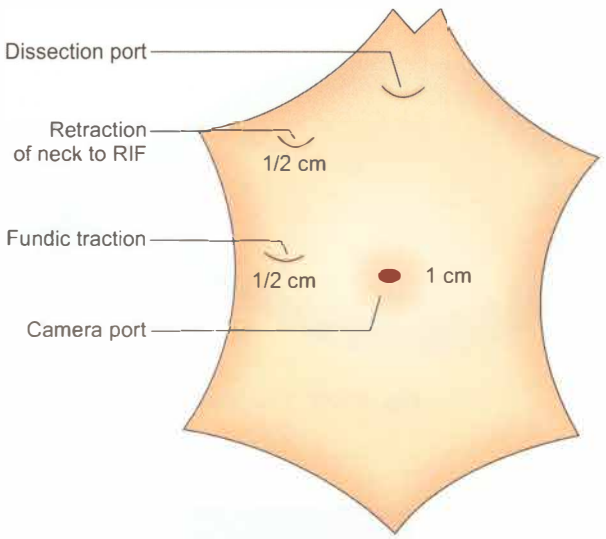


Fig. 25.35: Incisions for laparoscopic cholecystectomy

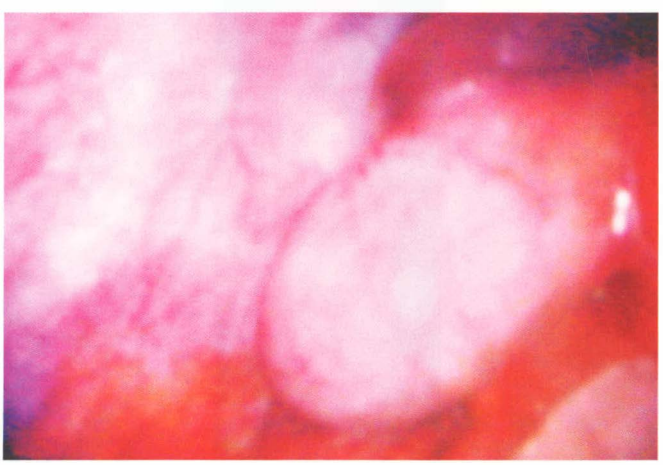


Fig. 25.36: Laparoscopic view of gall bladder

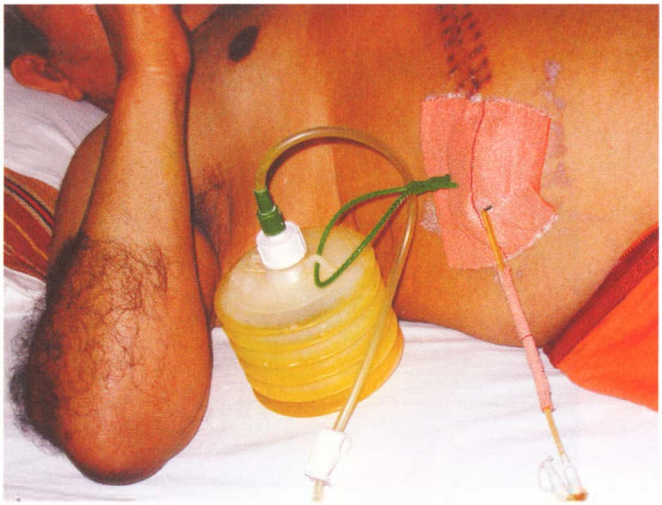


Fig. 25.37: Partial cholecystectomy could be done in this patient and all stones were cleared—asymptomatic since 3 years. It is a safe alternative to complete cholecystectomy. It is not a failure

- Following this, a laparoscope is introduced and a camera is attached. Three small, 1 cm incisions are made in epigastrium and 1/2 cm in the right hypochondrium. These are used for suction, instrumentation, cauterisation, dissection, retraction purpose—dissection port.
- Cystic duct and cystic artery are clipped and gall bladder is removed using gall bladder-holding forceps and is brought outside through the epigastric port (Figs 25.38 to 25.40).
- Bleeding from liver is controlled using lasers/cautery.
- The procedure is done under general anaesthesia. It may take 1–3 hours depending upon the experience of the surgeon.

Advantages

1. Hospital stay is 1–2 days, recovery is very fast.
2. Pain is minimal. Hence, mobilisation of the patient is much better and easy.
3. It gives an acceptable and better cosmetic result.
4. Complications such as adhesions and incisional hernia are rare.

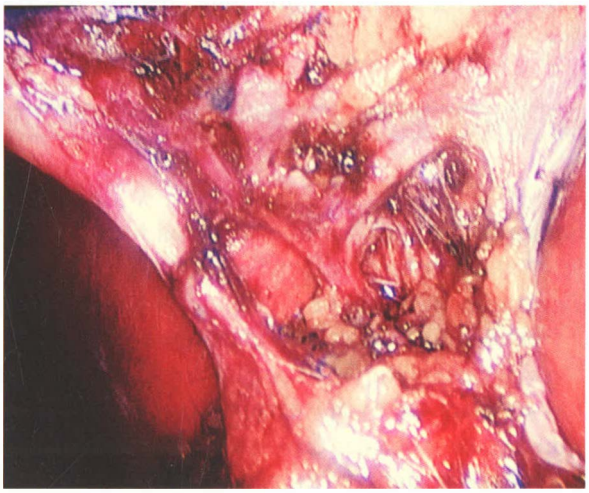


Fig. 25.38A: Cystic artery and cystic duct are identified and separated

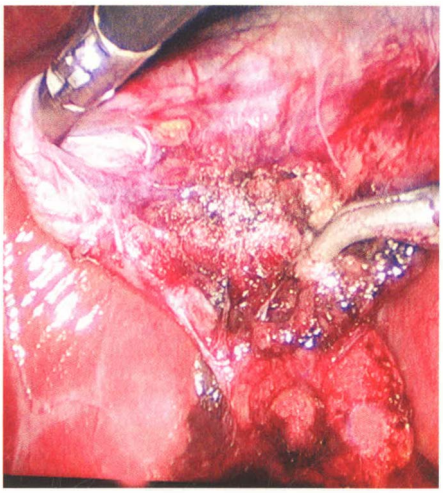


Fig. 25.38B: Cystic artery is isolated

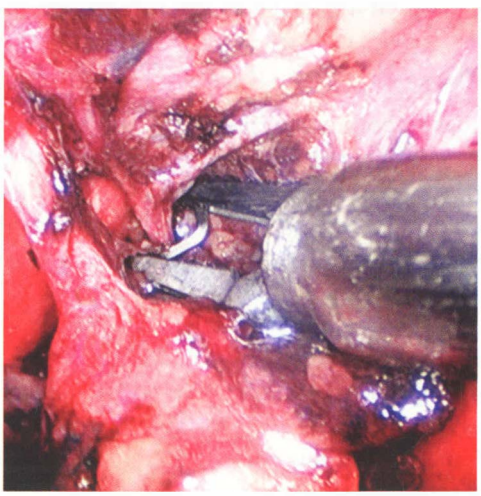


Fig. 25.39A: Cystic artery is being clipped

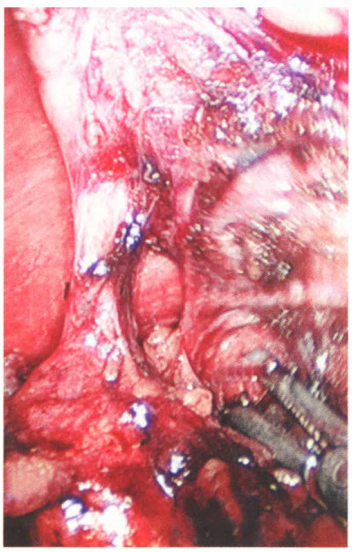


Fig. 25.39B: Cystic artery is divided

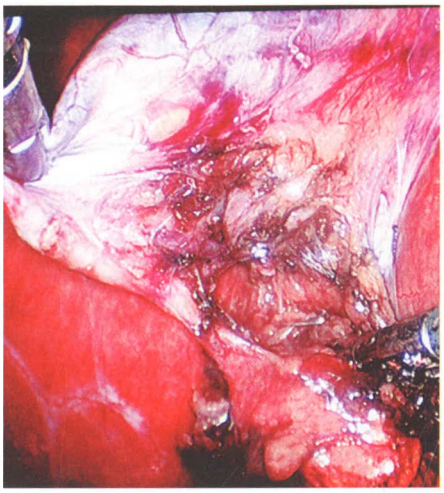


Fig. 25.40A: Calot's triangle is completely dissected

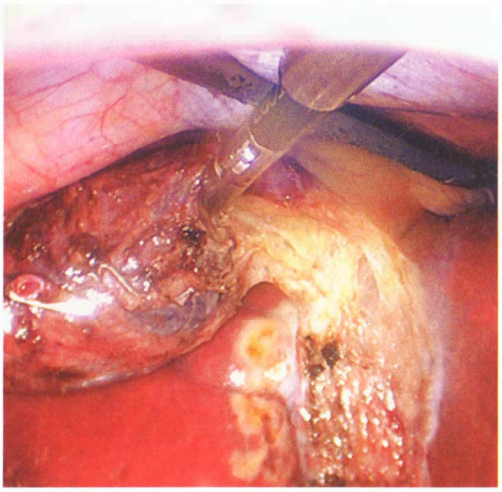


Fig. 25.40B: Gall bladder is mobilised from liver bed

MEDICAL TREATMENT OF GALL STONES

It is indicated for **pure cholesterol** stones only.

Patient selection

- Patients with functioning gall bladder proven by OCG or scintigraphy.
- Young, thin, female patients.
- Tiny (< 5 mm), translucent, floating stones

Drawbacks

- Recurrence of stones once treatment is stopped
- Life-long maintenance is needed.
- After dissolution of the stones, lithotripsy or extracorporeal shock wave lithotripsy should be done.

Types of medical treatment

1. Oral dissolution treatment

- **Drugs used:** CDCA—chenodeoxycholic acid
  UDCA—ursodeoxycholic acid
- **Mechanism of action:** They inhibit HMG-CoA, a rate limiting step in the synthesis of cholesterol, thereby increasing the bile salt pool. UDCA also acts by decreasing cholesterol absorption in GIT.

2. Direct contact dissolution

MTBE—methyl terbutyl ether is the drug which is given through a catheter placed in gall bladder percutaneously.

Drawbacks: Explosive and toxic if it enters bile duct or duodenum.

Indications

- High-risk patients with symptomatic stones, refusing surgery.
- The patient must have patent cystic duct (proven by OCG/ scintigraphy).

Side effects: Haemorrhage and catheter displacement.

Thus we have completed gall stones and their complications in the gall bladder. Now we will study the complication of gall stones in the common bile duct, mainly obstructive jaundice.

OBSTRUCTIVE JAUNDICE (SURGICAL JAUNDICE)

Definition

- Jaundice that occurs due to obstruction to the outflow of bile is called obstructive jaundice.
- Since these cases have to be managed by surgical intervention, it is also called surgical jaundice.
- However, haemolytic jaundice cases are not obstructive but a few are managed by splenectomy (surgical).
- Before we start a detailed discussion of obstructive jaundice, we will study anatomy of the pancreas. This is an important long case in the university examination. For a better understanding of obstructive jaundice, **students are requested to understand various causes of obstructive jaundice which are given in the subsequent pages.**

SURGICAL ANATOMY OF THE PANCREAS

The pancreas is both endocrine and exocrine organ situated retroperitoneally behind the stomach. It is a soft and fleshy gland (pancreas—all flesh), extending from the duodenum on the right side to the spleen on the left side, the entire length being 6 inches (Fig. 25.41). It weighs approximately 80 g.

Parts

- The head lies within the C-loop of duodenum. The uncinate process projects from the left inferior portion of the head over which course the superior mesenteric vessels. There are 5–6 small thin veins connecting this portion of the head with superior mesenteric veins. These veins have to be carefully divided during pancreaticoduodenectomy.

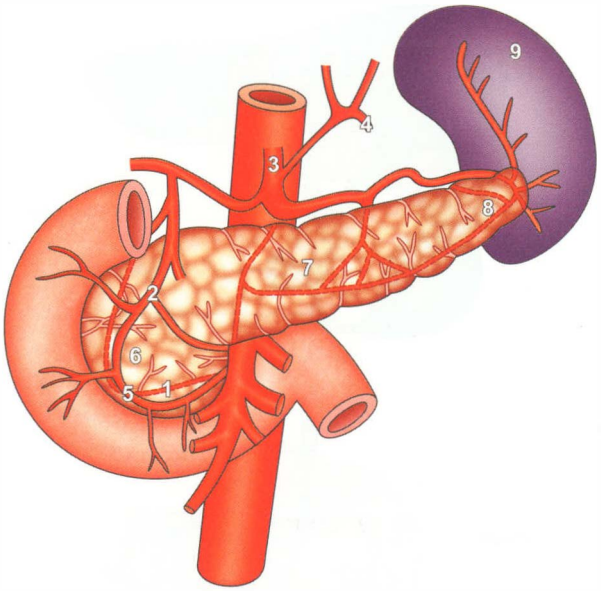


Fig. 25.41: Blood supply and parts of pancreas: (1) Uncinate process, (2) Superior pancreaticoduodenal, (3) Coeliac artery, (4) Splenic artery, (5) Inferior pancreaticoduodenal, (6) Head of pancreas, (7) Body of pancreas, (8) Tail of pancreas and (9) Spleen

Superior mesenteric vein continues above as portal vein after joining the splenic vein. During pancreaticoduodenectomy for periampullary carcinoma, infiltration into the portal vein should be ruled out before any major structure is divided. This is done by inserting a finger between the portal vein and head of pancreas, both from above and below.

- The neck is about 2 cm and is related posteriorly to superior mesenteric vessels.
- Body and tail: The head and neck continue as body which is placed transversely. It slopes upwards across the aorta and ends as tail of the pancreas, which is enclosed within lienorenal ligament along with splenic vessels. A large cystadenoma arising from the tail of the pancreas can move with respiration because of its contact with the spleen.

Blood supply of the pancreas

Arterial supply (Fig. 25.41)

- Splenic artery is the chief artery supplying the neck, body and the tail. Arteria pancreatica magna refers to one large branch of splenic artery.
- Superior and inferior pancreaticoduodenal arteries supply not only head of pancreas but also the adjacent duodenum. Thus, during any surgery which involves excision of the head, the C-loop of the duodenum is also removed. Thus, pancreaticoduodenectomy becomes a major surgery.

Venous drainage

- Body, neck and tail drain into splenic vein by means of multiple small veins.
- The head is drained by superior pancreaticoduodenal vein which drains into portal vein and inferior pancreaticoduodenal vein drains into superior mesenteric vein.

Islets of Langerhans (endocrine)

- 75% : β Cells
- 20% : α Cells
- 4% : δ Cells

Pancreatic duct (*see* page 608)

- The main pancreatic duct (duct of Wirsung), a tubular structure drains entire pancreas from tail to the head. It joins the common bile duct and forms ampulla of Vater. This ampulla opens on the duodenal papilla (a nipple-like elevation) in the 2nd part of the duodenum. Normal diameter of pancreatic duct is 2–3 mm. When it is dilated more than 6–8 mm, as in chronic pancreatitis, longitudinal pancreaticojejunostomy can be done.
- Accessory pancreatic duct of Santorini drains the uncinate process and lower portion of the head and opens into the duodenum 2 cm above the opening of the main duct. The two ducts communicate with each other at many sites.
- The main pancreatic duct is lined by columnar epithelium which becomes cuboidal in the ductules.

PANCREATIC JUICE

- Bicarbonate rich, protein rich, alkaline fluid
- 2.5 litres/day
- Rich in proteins 15 g of protein/day
- Thus acidic chyme in the duodenum is alkalinised by pancreatic juice
- Protein is secreted by acinar cells, fluid and electrolytes by ductal cells
- Secretions are stimulated by 3 phases
  a. Cephalic phase is mediated by acetylcholine—10% of secretion
  b. Gastric phase—mediated by gastrin and vagus—15%
  c. Intestinal phase—75% release mediated by release of secretion from duodenal acidification and stimulated release of bile into 2nd part of duodenum following entry of fat and proteins.

A CASE OF OBSTRUCTIVE JAUNDICE

Choledocholithiasis and carcinoma head of pancreas periampullary region are two important causes of obstructive jaundice. They constitute more than 90% of cases of obstructive jaundice treated surgically. Presence of jaundice, high-coloured urine, clay-coloured stools and itching gives the clue to the diagnosis. Palpable liver, palpable mass (cholangiocarcinoma, carcinoma head of pancreas) and palpable gall bladder (periampullary carcinoma) supports the diagnosis. Early diagnosis by imaging, endoscopy, ERCP, MRCP is possible. Since causes are many, they are treated

accordingly. Relief of pain, relief of jaundice with or without resection (in malignancies) should be done early to prevent complications such as cholangitis, liver abscesses, septicaemia, renal failure and even death.

AETIOLOGY (Fig. 25.42)

I. Causes in the lumen

1. Stones in the common bile duct
2. Ova, cysts, ascaris worms
3. Hydatid cyst of the biliary tree
4. Stone in the pancreatic duct/CBD junction

II. Causes in the wall

1. Periampullary carcinoma
2. Bile duct stricture
3. Stenosis of sphincter of Oddi (papillary stenosis)
4. Klatskin's tumour—carcinoma of the bile duct where right and left ducts join
5. Choledochal cyst
6. Post laparoscopic cholecystectomy

III. Causes from outside (due to pressure)

1. Carcinoma head of pancreas
2. Chronic pancreatitis
3. Lymph nodes at the porta hepatis obstructing the biliary tree.

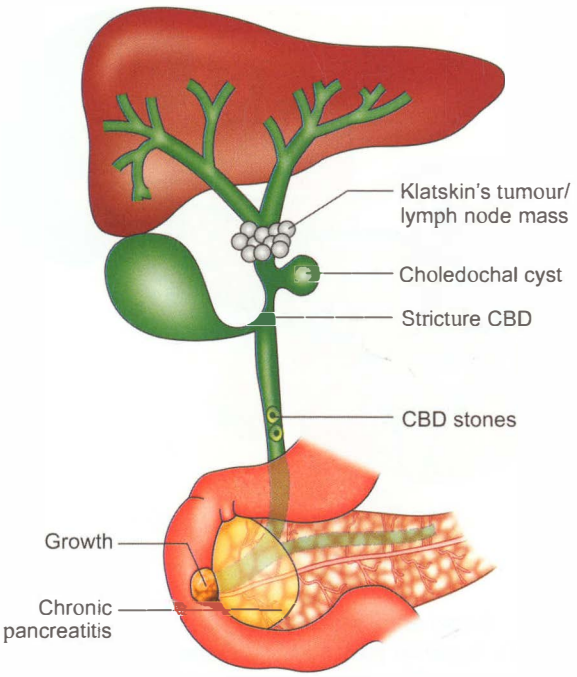


Fig. 25.42: Various causes of obstructive jaundice

CLINICAL FEATURES

First study to differentiate choledocholithiasis from carcinoma periampullary/head of pancreas (Table 25.1).

Table 25.1 Differences between stone in the CBD and periampullary carcinoma (Ca)/Ca head of pancreas

	Stone in the CBD	Carcinoma periampullary/head of pancreas
1. Age	30–50 years	50–70 years
2. Sex	More common in females	Equally common in both sexes
3. Duration of symptoms	Long duration	Short duration (1–3 months)
4. Symptoms		
Pain	It is due to a stone blocking the CBD resulting in spasm of CBD. It is severe colicky pain like gall stone colic.	There may be some discomfort in abdomen but colicky pain is not a feature. Pain is relatively rare in carcinoma head of the pancreas.
Fever	As a result of obstruction, multiplication of organisms results in fever.	When obstruction becomes severe, there is bile stasis. Cholangitis, fever with chills and rigors can occur.
Jaundice	Occurs due to obstruction. Once inflammation subsides all these 3 symptoms are relieved partly but they occur after sometime. Hence, intermittent pain, intermittent fever, intermittent jaundice are classical of a stone in CBD—**Charcot's triad**.	As a result of slow developing obstruction in periampullary region, jaundice is **persistent, progressive, painless, pruritic**. In 5% of cases, growth may ulcerate into the duodenum. It can cause melaena and jaundice may temporarily subside.
Stools	Since the obstruction is never complete, clay-coloured stools are not commonly seen.	Clay-coloured stools are common and when mixed with blood, it is called **silvery stools** or aluminium paint stools.
Pruritus	May be present but mild	Severe—due to bile salts in the circulation.
Loss of appetite	No	Significant
Loss of weight	No	Significant
5. Signs		
Jaundice	Deep yellow	Sometimes, greenish yellow
Anaemia	Absent	It is usually present
6. Per abdomen	Liver can be enlarged due to back pressure. It is smooth, with round border, firm in consistency.	Liver can be enlarged due to back pressure. If it is nodular, with sharp border, hard in consistency, it is due to secondaries in the liver.
7. Gall bladder	As a rule, gall bladder is not palpable	Gall bladder is palpable in 70–75% cases.
8. Metastasis	No	Left supraclavicular node, ascites, etc. may be seen.

Courvoisier's law (Figs 25.43 and 25.44)

- In a jaundiced patient, if the gall bladder is palpably enlarged, it is not due to stones (Key Box 25.10). In case of stones, previous inflammation would have made gall bladder fibrotic and hence, will not be palpable.
- From clinical point of view, 90% of cases of obstructive jaundice are due to stones, periampullary carcinoma or carcinoma of the head of pancreas.

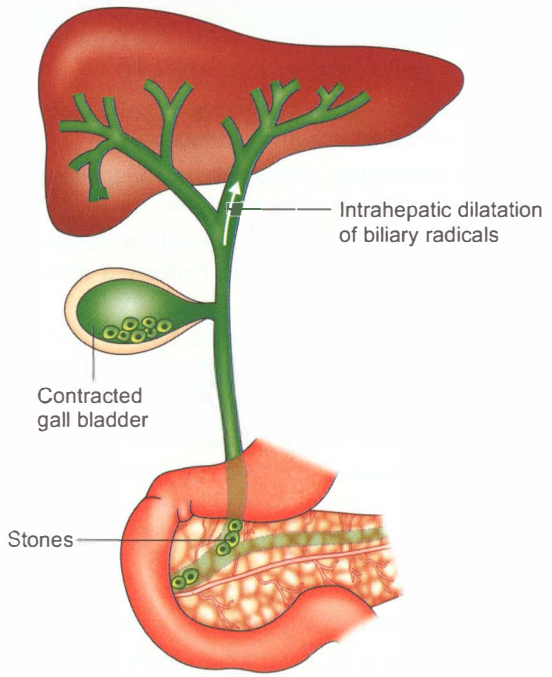


Fig. 25.43: Contracted thick gall bladder due to stones. Hence, not palpable

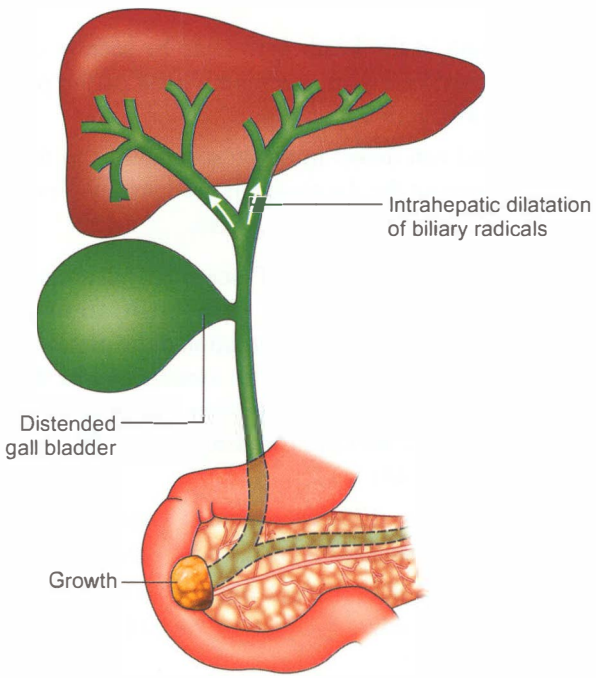


Fig. 25.44: Enlarged palpable gall bladder in cases of periampullary carcinoma

KEY BOX 25.10

CLINICAL FEATURES OF GALL BLADDER MASS

- Egg-shaped mass/pyriform shape
- Moves with respiration
- Tensely cystic, feels firm, tender and sometimes located in the right hypochondrium
- Superficially placed
- Intra-abdominal, intraperitoneal

Exceptions to Courvoisier's law

1. Double impaction: One stone in the CBD and one stone in the cystic duct.
2. Periampullary carcinoma in a patient who has undergone cholecystectomy.
3. Primary oriental cholangiohepatitis causing stones in the CBD (gall bladder is normal in these cases).

CHOLEDOCHOLITHIASIS (Table 25.1)

Types of CBD stones

I. Primary stones: These stones are formed in the CBD or within intrahepatic ducts. They are multiple, pigment stones or often mixed stones (Key Box 25.11). Various causes are:

1. Infections of biliary tree and infestation—parasites such as clonorchiasis.
2. Congenital—Caroli's disease or choledochal cyst.
3. Biliary dyskinesia—defective pathophysiology of biliary tree.
4. Other causes—diabetes, malnutrition.

II. Secondary stones: These stones originate from gall bladder and stay in CBD—usually supraduodenal portion—then get enlarged to attain large size over a period of time.

These stones can give rise to cholangitis.

Cholangitis

Bacterial infection of bile duct is called cholangitis.

KEY BOX 25.11

CBD STONES

- Only 4–12% of all CBD stones
- These stones do form primarily in common bile duct
- Almost all primary CBD stones are pigment type (brown stones).
- Associated with bile duct stasis and infection (bacteria)
- Soft, easily crumble when manipulated
- Associated with biliary stricture, papillary stenosis or sphincter of Oddi dysfunction.
- Mechanism; Stasis—bacteria secrete bacterial glucuronidase which causes deconjugation of bilirubin diglucuronide. Bilirubin gets precipitated as calcium salt.

Predominant organisms

- *E. coli,* Klebsiella, Pseudomonas, Enterococci, Proteus.
- Bacteroides and other anaerobes (*Clostridium perfringens*)

Important causes: Cholangitis.

- Choledocholithiasis
- Biliary stricture
- Neoplasm
- Less important causes are pseudocyst of pancreas, chronic pancreatitis stenosing sphincter of Oddi, biliary parasitic infections, etc.
- Post ERCP, if stent cannot be passed as in obstructive jaundice cases.

Symptoms and signs

- Biliary colic, jaundice and chills and rigors are called Charcot's triad.
- Tenderness may be present in the upper abdomen.

Investigations

- Leukocytosis, high bilirubin levels and alkaline phosphatase levels are diagnostic tests.
- Ultrasound, CT scans, ERCP are indicated to confirm/rule out the various causes.

Treatment

- Intravenous antibiotics
- Emergency endoscopic sphincterotomy, extraction of stones in choledocholithiasis, endoscopic stoning in cases of stenosis or stricture.
- Percutaneous transhepatic biliary drainage (PTBD) in high obstructions.
- Laparotomy—drainage of CBD—T-tube insertion.

Reynold's pentad of acute obstructive cholangitis

Few cases of CBD stones present with serious problems of cholangitis described as Reynold's pentad

1. Persistent pain
2. Fever
3. Persistent jaundice
4. Shock
5. Altered mental status

INVESTIGATIONS IN OBSTRUCTIVE JAUNDICE

1. **Hb%** is low in malignancy.
2. **TC, DC** are increased in case of infections.
3. **BT, CT, PT** are altered in case of obstructive jaundice.
4. **Urine** for urobilinogen is negative in obstructive jaundice.
5. **Serum alkaline phosphatase:** Normal value 60–300 units/L More than 500 units is suggestive of obstructive jaundice
   - These are the enzymes which bring **hydrolysis** o phosphate esters in alkaline medium.
   - Sources of alkaline phosphatase include **liver, biliary tree**, bone, intestine, kidney.
   - Excretion is mainly through biliary tree (Key Box 25.12)

KEY BOX 25.12

ALKALINE PHOSPHATASE

- It is the product of epithelial cells of cholangioles—↑ levels are due to increased enzyme production.
- Intrahepatic cholestasis, cholangitis, extrahepatic obstruction are the chief factors causing elevation.
- Focal lesions in the liver—single hepatic metastasis or liver abscess or a tumour can cause increased levels without jaundice.
- In cholangitis, bilirubin may be normal but alkaline phosphatase may be very high.

Gross elevation	Mild elevation
Obstructive jaundice, biliary cirrhosis, bone disease	Metastasis in the liver hepatic abscess hepatitis

6. **Abdominal ultrasound:** It is the **most useful, noninvasive, reliable and quick investigation for obstructive** jaundice. Dilated biliary radicles, both intrahepatic and extrahepatic can be demonstrated (First clue in obstructive jaundice).
   - Stones can be diagnosed with their posterior acoustic shadow.
   - Mass lesion in the head region can be seen in cases of chronic pancreatitis or carcinoma head of the pancreas causing obstructive jaundice.
   - Ultrasound can detect multiple secondaries in the liver, thus, favouring the diagnosis of malignancy. Endosonogram can detect lymph nodes also.
7. **CECT (contrast enhanced CT) scan:** A head mass of even 2–3 cm in size and portal vein infiltration can be demonstrated by CT scan. Obliteration of fat plane between the mass and superior mesenteric vessels can be demonstrated by CT scan which decides the operability of periampullary carcinoma or carcinoma head of the pancreas (Figs 25.45 to 25.48).
   - CT scan cannot differentiate head mass of carcinoma from chronic pancreatitis (PET scan may differentiate).
   - CT scan can also detect coeliac nodes, presence of which is a contraindication for radical resections (Key Box 25.13).
   - Take precautions against contrast induced nephropathy.

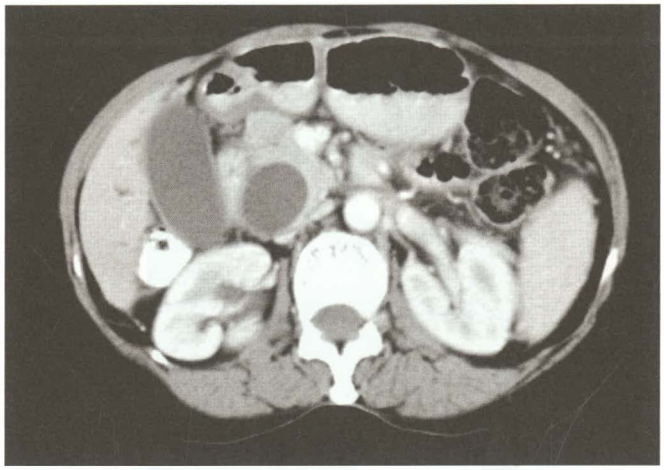


Fig. 25.45: CT scan showing hugely distended gall bladder and common bile duct—periampullary carcinoma. Growth was resectable. He underwent **Whipple's operation**

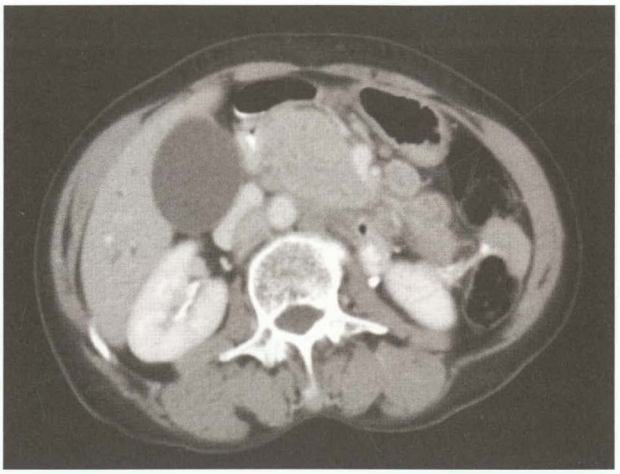


Fig. 25.46: Large 6 cm mass in the head of pancreas—patient not only had obstructive jaundice but also duodenal obstruction. He underwent **triple bypass**

Fig. 25.47: 6 cm mass abutting mesenteric vessels and involving root of the mesentery. Such masses are usually nonresectable

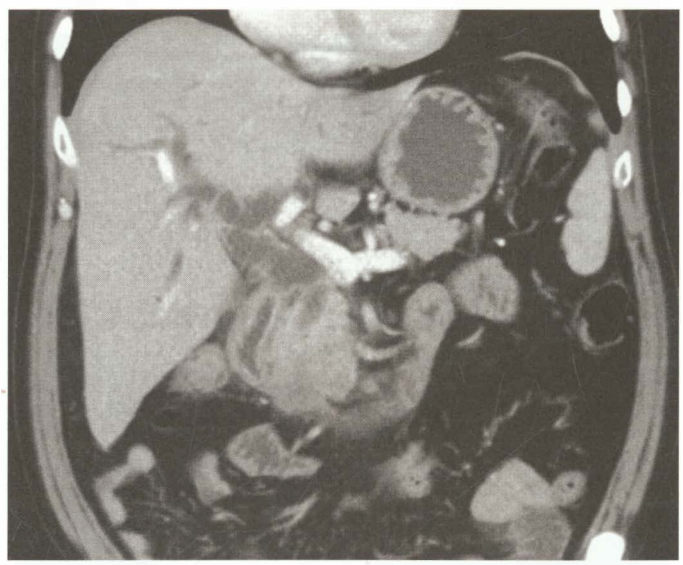


Fig. 25.48: Large head mass with obstructive jaundice with intrahepatic dilatation. When in doubt, diagnostic laparoscopy will help—to operate or palliate nonsurgically

KEY BOX 25.13

COMPLICATIONS OF PERCUTANEOUS CT-GUIDED BIOPSY OF HEAD MASS

Death
Enzymatic autodigestion
Abscess
Traumatic fistula
Haemorrhage
Remember as **DEATH**

8. **Endoscopy** is useful to diagnose a periampullary carcinoma which may be seen as an ulcerative lesion in the second part of the duodenum. Biopsy can also be taken which shows adenocarcinoma (Fig. 25.49).
   - In case of obstructive jaundice due to stones, smooth bulge can be seen in the second part of the duodenum. In carcinoma, ulcerated lesion can be seen.
9. **Barium meal** follow through to see the C-loop of duodenum (Fig. 25.50).
   - In periampullary carcinoma, there may be distortion of the medial border of the duodenum giving rise to **Inverted 3 sign**.
   - In carcinoma head of the pancreas there may be widening of C-loop of duodenum—**Pad sign**.
10. **ERCP (endoscopic retrograde cholangio-pancreatography)** (Figs 25.51 to 25.53)
    - With the help of a side viewing endoscope, ampulla of Vater is cannulated and a radio-opaque dye is injected.
    - It fills up the biliary and pancreatic system.

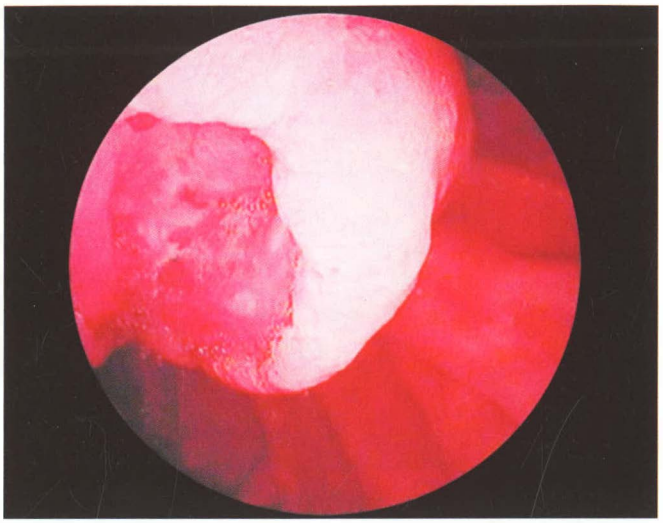


Fig. 25.49: Endoscopy showing growth in the periampullary region. These lesions are usually resectable

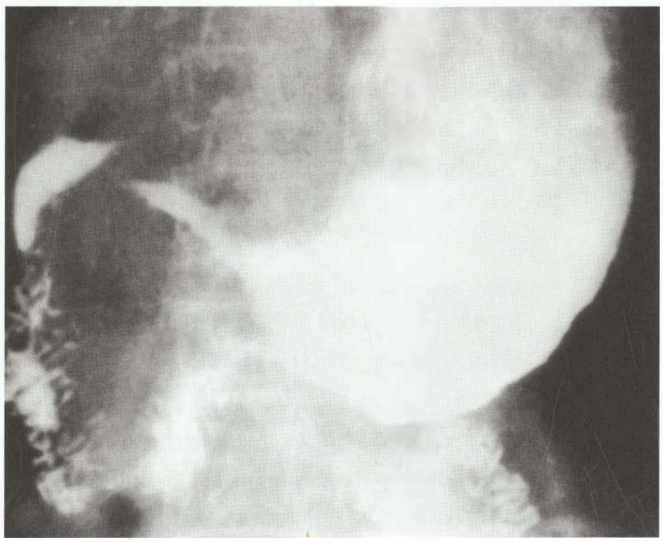


Fig. 25.50: Barium meal showing widening of C-loop of duodenum. **It is not done nowadays**

Interpretation

- Stones appear as filling defects in the CBD or in the common hepatic duct (CHD) (Figs 25.51 to 25.53), which may be mobile (change position if patient is moved).
- A periampullary carcinoma gives rise to an irregular filling defect or there may be total cut off in the flow of dye.
- Chronic pancreatitis may show the dilated duct and stones in the pancreatic duct—***'chain of lakes' appearance***.

Uses

If stones in the CBD are diagnosed, they can be treated in the following ways:

- Extraction by using a basket.
- Large stone can be crushed by using a lithotripter and can be extracted.

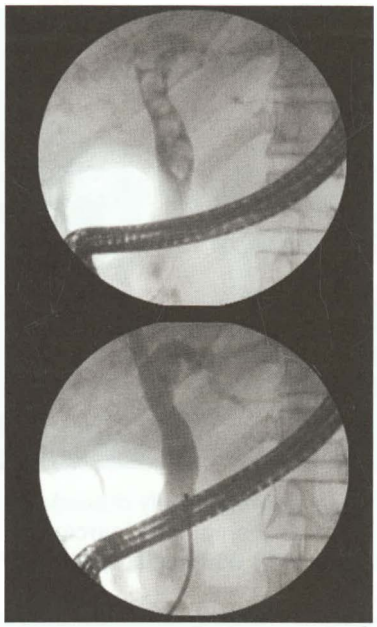


Fig. 25.51: ERCP showing stones in the CBD, stones are extracted and stent has been placed in the common bile duct

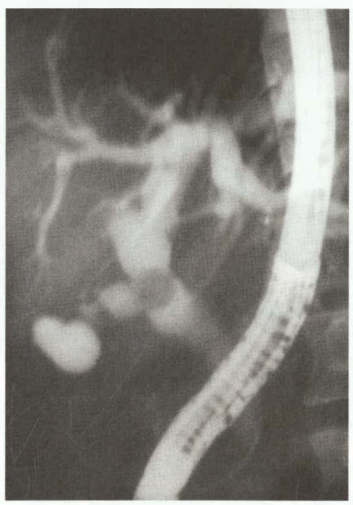


Fig. 25.52: ERCP showing stones in the CBD and contracted gall bladder

Fig. 25.53: ERCP showing stones in the CBD and contracted gall bladder

- Sphincterotomy (incision of sphincter of Oddi) can be done to facilitate extrusion of small stones.
- In patients with cholangitis with obstructive jaundice, stenting of common bile duct can be done to relieve obstruction. **Stent removal is necessary** at a later date.
- In selected patients with **biliary strictures**, stent is placed after ERCP to relieve obstructive jaundice (sometimes permanent in malignancies).
- In selected patients with **chronic pancreatitis**, pancreatic duct can be stented to relieve pain.

PEARLS OF WISDOM

ERCP and stenting is NOT indicated in operable cases of carcinoma periampullary/head of pancreas unless cholangitis is present.

Complications

Severe infection of biliary tree (cholangitis) and acute pancreatitis can occur in 1–2% of the patients. Hence, prophylactic antibiotics are given before the procedure.

11. **Endosonogram:** Endoscopy aided ultrasound can detect missed stones in the CBD. It can also detect pancreatic head mass, lymph nodes. Endosonoguided FNAC can also be done (Fig. 25.54).

12. **Percutaneous transhepatic cholangiography (PTC):** Using an ultrasound image-intensifier, a dilated biliary radicle is identified within the liver and a fine needle (Chiba needle[1]) is introduced into it. The stylet is then removed and a radio-opaque dye injected. Chiba needle is 15 cm long and 0.7 mm in diameter (Figs 25.55A and B, Key Box 25.14).

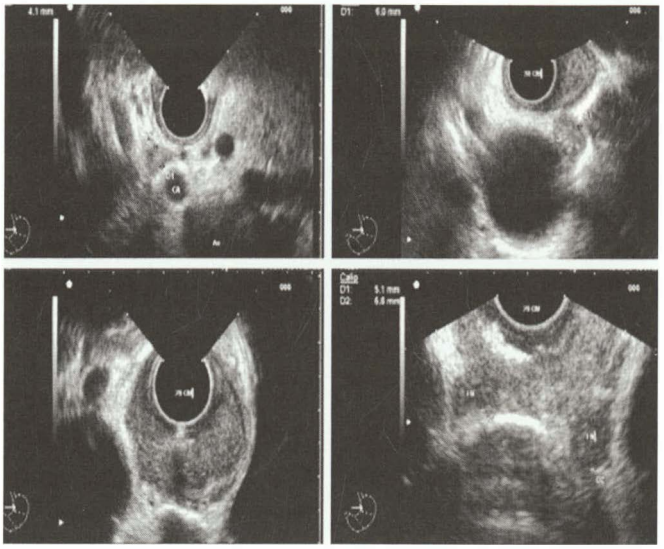


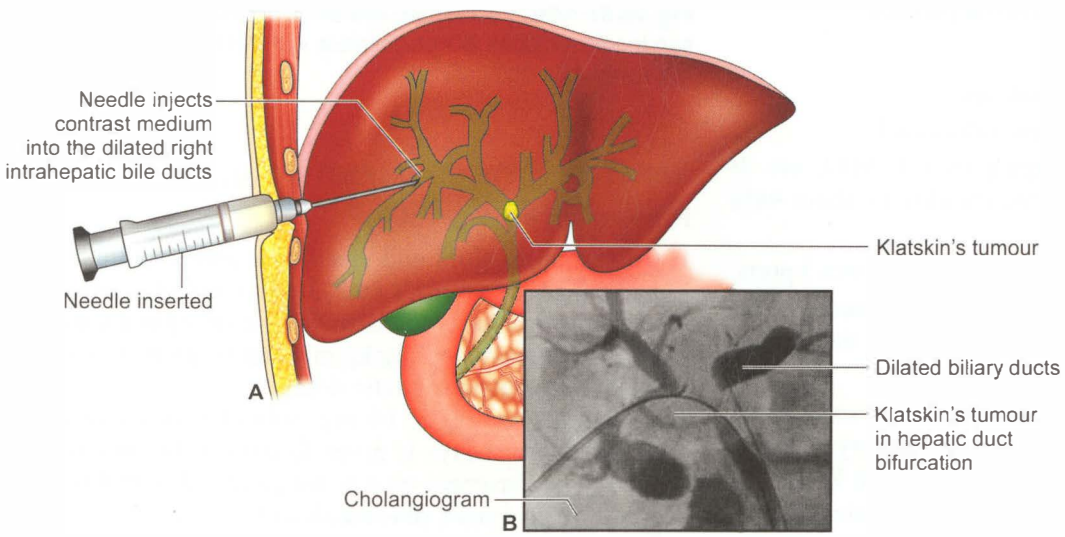
Fig. 25.54: Endosonography showing pancreatic tumour with coelic lymph nodes (*Courtesy:* Dr Ganesh Pai, HOD, Medical Gastroenterology, KMC, Manipal)

Precautions

- BT, CT, PT should be normal. Otherwise, vitamin K injection 10 mg is given IV or SC for 3 days.
- If there is a bleeding tendency, this procedure should not be done.
- Broad spectrum antibiotics are given before the procedure.

Complications

- Infection, cholangitis, septicaemia
- Biliary leak can be significant producing abdominal pain and guarding. Hence, PTC should be done just prior to the surgery.
- Haemorrhage



Figs 25.55A and B: (A) Diagrammatic representation of percutaneous transhepatic cholangiography (PTC); (B) Dye had failed to fill-up the bifurcation of the common hepatic duct suggesting the obstruction

[1]Chiba: It is the name of a university in Japan.

Fig. 25.55C: PTBD catheter inside CBD

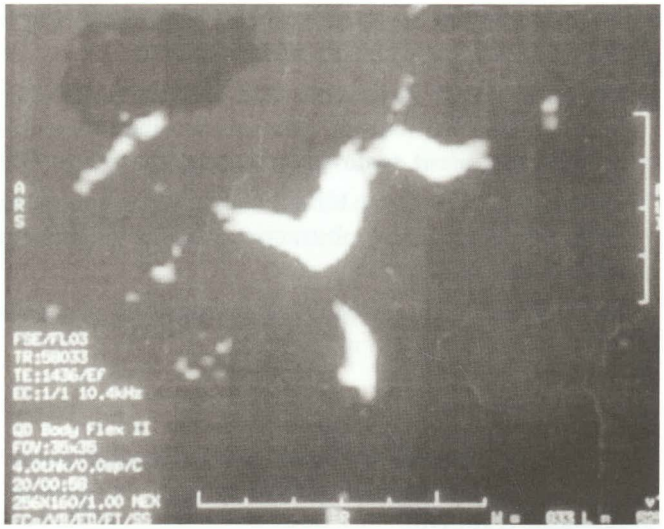


Fig. 25.56: MRC (magnetic resonance cholangiogram) showing high stricture—possibility of Klatskin's tumour (cholangiocarcinoma)

KEY BOX 25.14

FACTS ABOUT PTC

1. In the diagnosis of high strictures, PTC is better than ERCP.
2. It can also be used in ERCP failure cases.
3. In the diagnosis of Klatskin's tumour, PTC is extremely useful. It can also delineate the dilated proximal duct, which helps in planning for a biliary-enteric anastomosis.
4. Catheter can be kept in the bile ducts to provide external drainage as in strictures or in inoperable malignancies with obstructive jaundice (Fig. 25.55C).

13. **MRI scan (MRCP)** (Figs 25.56 and 25.57)
    - It is the investigation of choice in cases of obstructive jaundice or of high strictures and cholangiocarcinomas.
    - It is noninvasive and delineates the bile ducts very well so that a biliary bypass can also be planned.
14. **Diagnostic laparoscopy**
    - It is used in many GI malignancies
    - It can be aided by laparoscopic ultrasound
    - It can complement the staging by CT, MRI, etc. It can improve prediction of resectability to about 98% accuracy.
    - It is a simple but invasive investigation, requires 3 ports.
    - Specially excellent to detect ***peritoneal metastasis*** (which cannot be picked by other tests) which is the sign of inoperability.
15. **CA 19–9:** Often head mass can be due to carcinoma/chronic pancreatitis. Biopsy is not mandatory. Clinical suspicion of a head mass may be treated with Whipple's pancreaticoduodenectomy. However, gross elevation of CA 19–9 (carbohydrate antigen), will suggest carcinoma. Figure 25.58 shows investigations and treatment of obstructive jaundice.

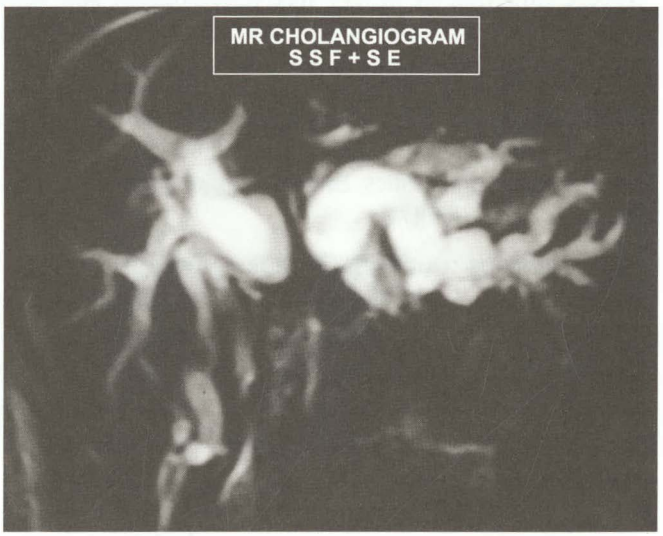


Fig. 25.57: MRC intrahepatic dilatation, another case of cholangiocarcinoma—biopsy is not possible with MRI. Brush cytology is possible while doing ERCP

TREATMENT OF OBSTRUCTIVE JAUNDICE

Preoperative preparation

1. Correction of fluid and electrolyte status and adequate hydration before surgery for 2–3 days is essential, especially when patients have vomiting or have developed sepsis.
2. Injection dopamine 2 µg/kg/min can be given to improve the urinary output (diuretic dose).
3. Injection vitamin K, 10 mg, subcutaneously or intravenously for 3 days is given to correct the prothrombin time. If prothrombin time is not corrected with this treatment, **fresh frozen plasma** should be given.
4. Broad spectrum antibiotics are given before, during and after surgery.
5. Adequate blood transfusion to correct anaemia.

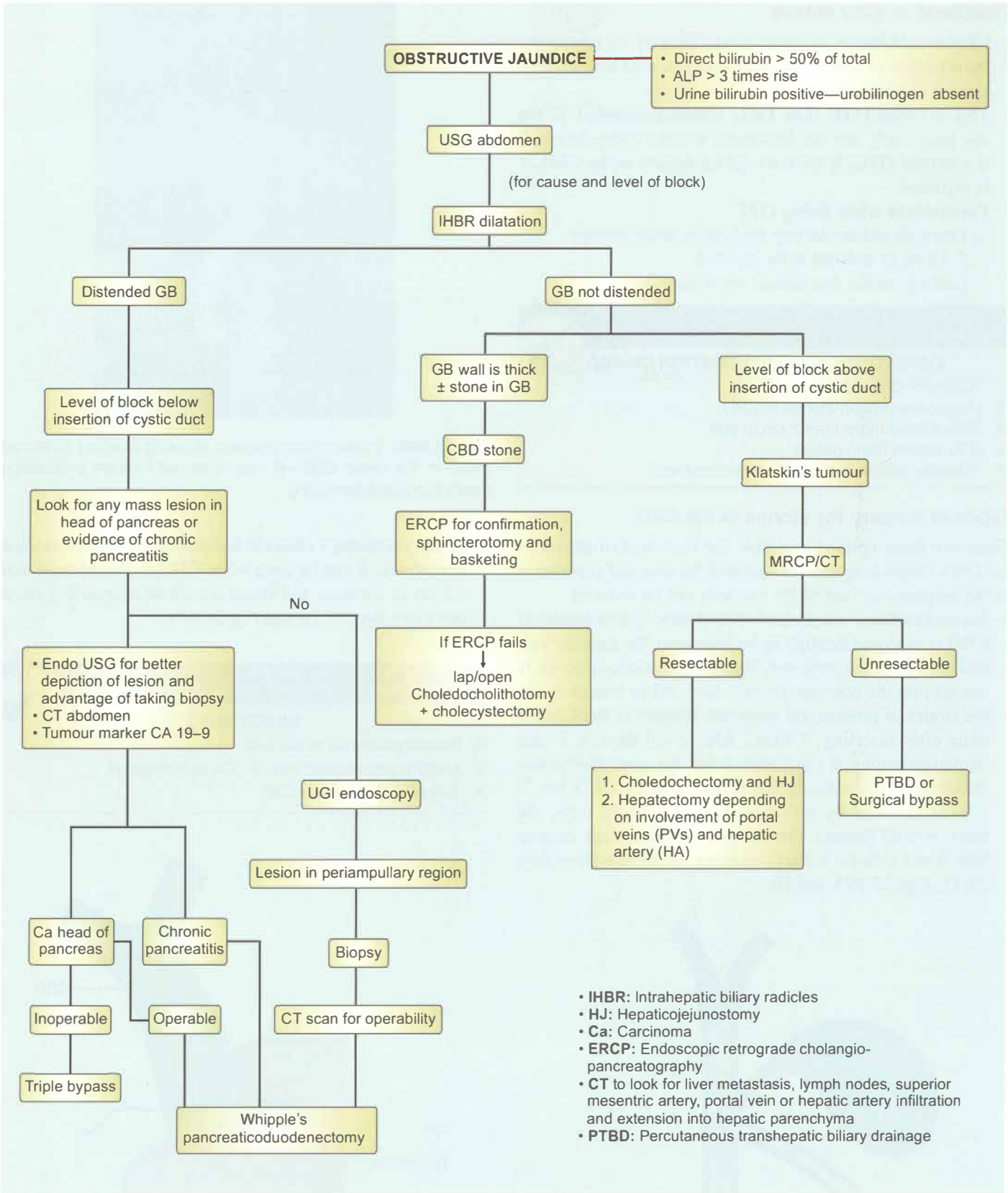


Fig. 25.58: Investigations and treatment of obstructive jaundice (*Courtesy:* Dr Ankur Sharma, Asst. Professor, Dept. of Surgery, KMC, Manipal)

Treatment of CBD stones

- Cholecystectomy is done first. This is followed by introduction of a cannula into the cystic duct and a radio-opaque dye is injected.
- This is called **OTC** (**O**n **T**able **C**holangiography). If the dye goes freely into the duodenum without filling defect, it is a normal OTC. If there are filling defects in the CBD, it is explored.
- **Precautions while doing OTC**
  - There should not be any air bubble in the syringe.
  - 5–10 ml of dye has to be injected.
  - Leakage of the dye should not occur.

KEY BOX 25.15

INDICATIONS FOR EXPLORATION OF CBD

1. History of Charcot's triad
2. Ultrasound proven stones in CBD
3. CBD dilated more than 1 cm in size
4. OTC shows filling defect
5. Palpable stones in the CBD (open method)

Types of surgery for stones in the CBD

There are three options available for treatment of stones in the CBD. Depending upon the merit of the case and experience of the surgeon any one of the methods can be selected.

1. ***Supraduodenal choledocholithotomy:*** Supraduodenal CBD is explored through an incision over the anterior wall and the stones are removed. Operating choledochoscope is passed into the common hepatic duct and its branches and the stones, if present, are removed. Closure of the CBD is done after inserting 'T-tube'. After 8–10 days, a T-tube cholangiography is done and if the dye goes freely into duodenum and no filling defect is seen in the CBD, the T-tube is removed by gentle traction. By 10–12 days, the track is well formed. Hence, even if minor leak occurs, bile flows outside without causing peritonitis (Key Box 25.15, Figs 25.59A and B).

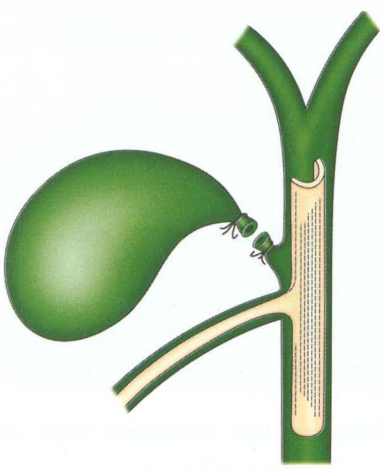


Fig. 25.59A: Cholecystectomy, CBD exploration followed by insertion of T-tube

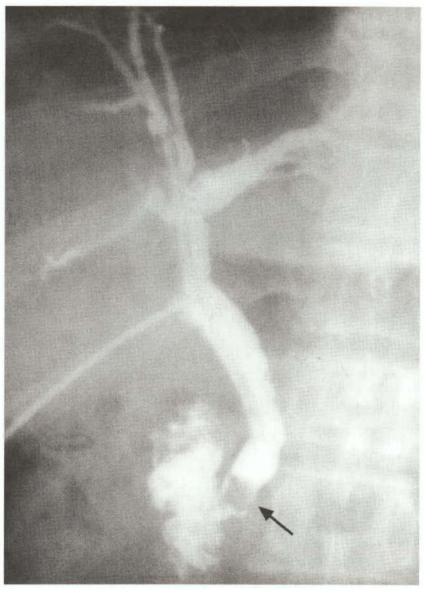


Fig. 25.59B: T-tube cholangiogram showing residual (retained) stone in the lower CBD—it was removed through endoscopic papillotomy and basketing

2. ***Cholecystectomy + choledocholithotomy + choledochoduodenostomy:*** It can be done when CBD is dilated more than 1.5 cm in diameter and stoma should be at least 2–3 cm in size (Key Box 25.16 and Fig. 25.60).

KEY BOX 25.16

INDICATIONS

- Recurrent stones in the CBD
- Multiple intrahepatic stones (Caroli's disease)
- Stricture of the lower CBD

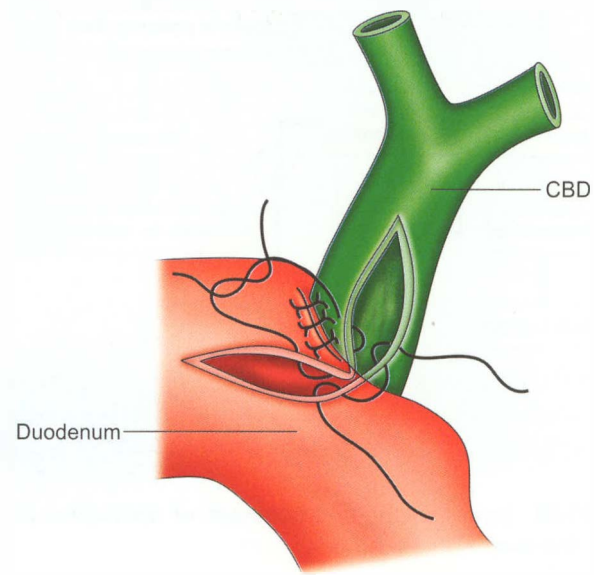


Fig. 25.60: Choledochoduodenostomy

Advantages of choledochoduodenostomy (Key Box 25.16)

- Biliary leak is negligible
- There is no worry even if there are retained stones in the CBD.
- It is a permanent solution for stenosis, stricture or multiple intrahepatic stones.

3. **Preoperative ERCP, sphincterotomy + extraction of stones followed by laparoscopic cholecystectomy**
   This method has become the choice today. Expertise and sophisticated equipment are necessary for this (Fig. 25.61).

Fig. 25.61: Stone was removed through endoscopic papillotomy followed by basketing. There is a small risk of perforation and pancreatitis in these patients (*Courtesy:* Dr Ganesh Pai, Prof and HOD of Medical Gastroenterology, KMC, Manipal)

Natural history of CBD stones (Key Box 25.17)

KEY BOX 25.17

NATURAL HISTORY OF CBD STONES

Symptom	*% of patients*
• Asymptomatic	— 40%
• Cholangitis	— 20%
• Jaundice	— 20%
• Biliary colic	— 15%
• Pancreatitis	— 5%

Treatment of periampullary carcinoma

I. Surgical treatment (Figs 25.62 to 25.69)

1. **Radical pancreaticoduodenectomy—'Whipple's operation'** (Figs 25.62 and 25.63).
   - In this operation, the growth along with 'C' loop of duodenum up to DJ flexure, proximal jejunum, head of the pancreas up to the neck are removed and partial gastrectomy is done. This is followed by:
   - Pancreaticojejunal anastomosis (PJ), gastrojejunostomy (GJ) and choledochojejunostomy (CJ).

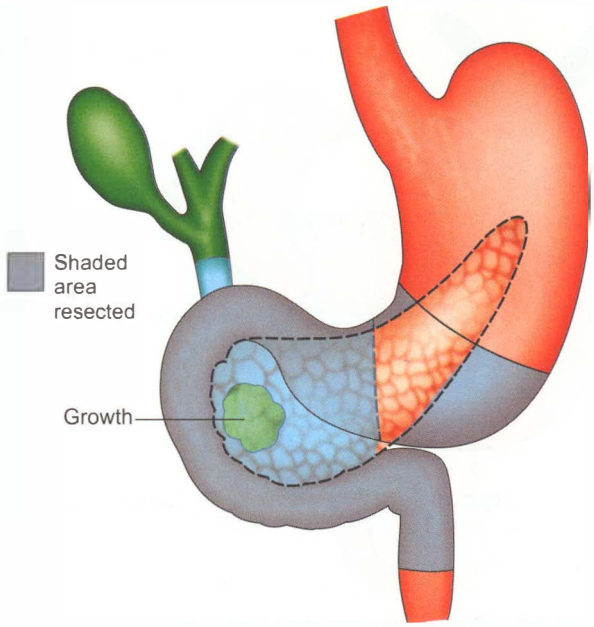


Fig. 25.62: Diagrammatic representation of Whipple's resection

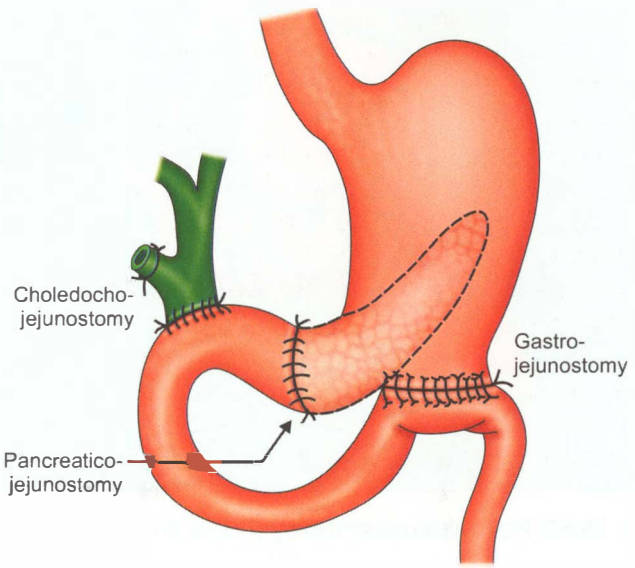


Fig. 25.63: Reconstruction following Whipple's operation

PEARLS OF WISDOM

Preoperative tissue diagnosis is NOT necessary with appropriate clinical and imaging findings, specially in head masses.

   - This is a major operation and carries 5–10% mortality due to pancreatic leakage or biliary leakage.
   - Whipple's operation is indicated in cases of mobile growth with no metastasis and where the general condition of the patients is reasonably good.

2. **Pylorus-preserving pancreaticoduodenectomy (PPPD)**
   In this operation, pylorus is preserved. Thus, gastric motility is not disturbed (Fig. 25.64).

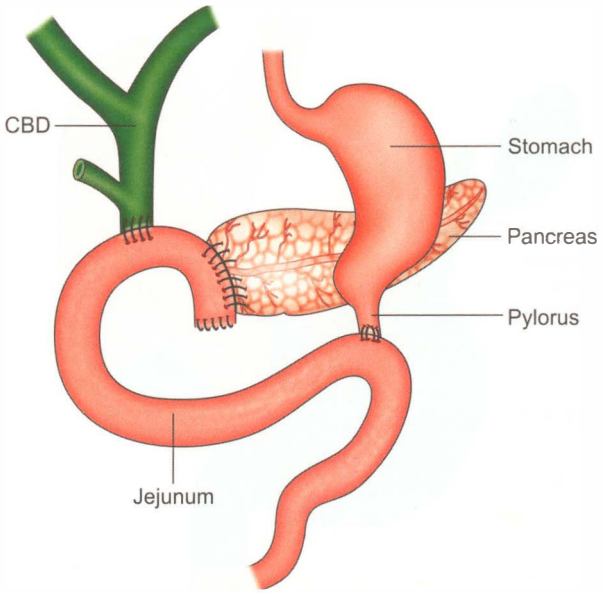


Fig. 25.64: Pylorus-preserving pancreaticoduodenectomy (PPPD)

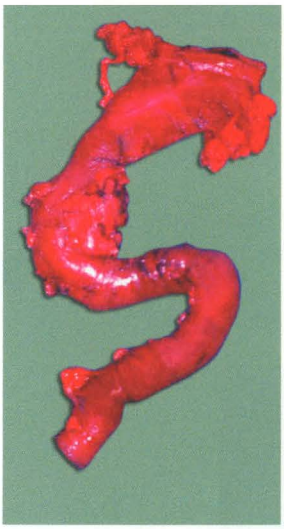


Fig. 25.65A: Whipple's resection specimen

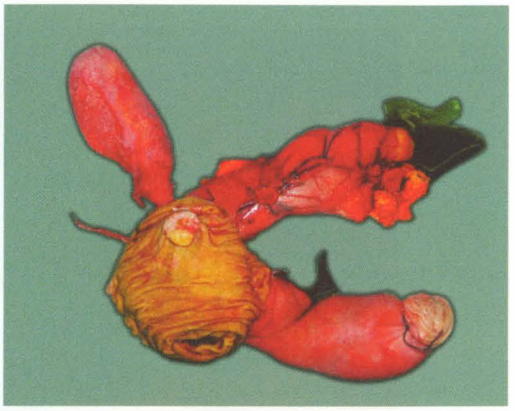


Fig. 25.65B: Whipple's specimen (*Courtesy:* Dr Satyanarayana N, Associate Professor, Yenepoya Medical College, Mangalore)

Fig. 25.66: Pancreaticogastrostomy (PG) is an alternative to PJ

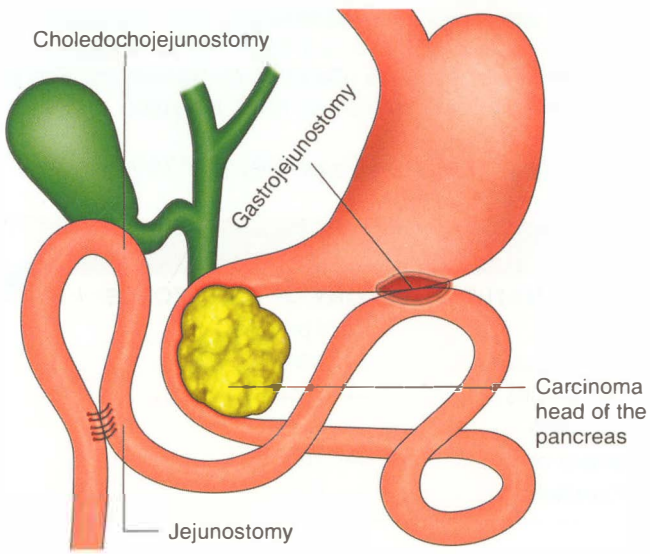


Fig. 25.67: Palliative triple bypass

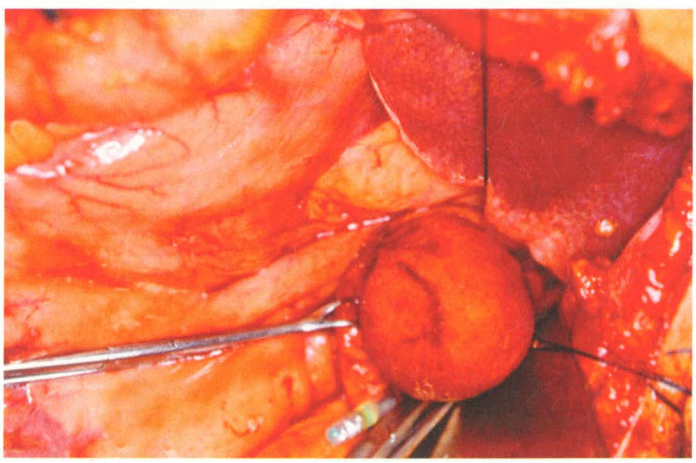


Fig. 25.68: Distended gall bladder ready for anastomosis

'TUNNEL OF LOVE'

During resectional surgery for pancreatic head and uncinate process tumours (Whipple's pancreaticoduodenectomy), after a preliminary search for metastases, a tunnel (the so-called tunnel of love) needs to be developed behind the neck of the pancreas and anterior to the underlying visceral vessels before concluding that the lesion can be resected.

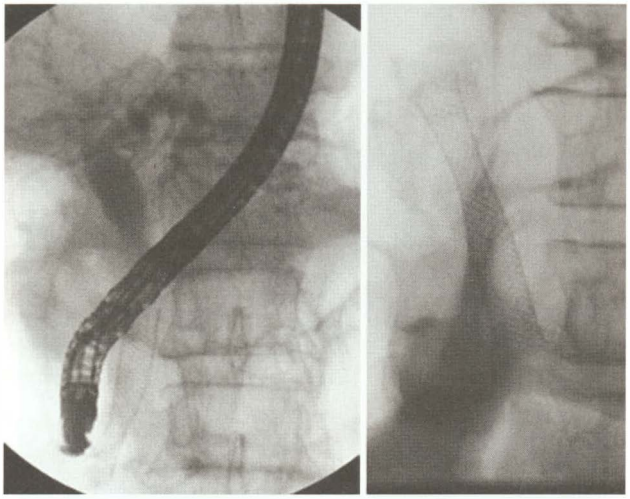


Fig. 25.69: Palliative stenting for carcinoma head of the pancreas

3. ***Triple bypass:*** **Cholecystojejunostomy + enteroenterostomy + gastrojejunostomy**
   - This is a palliative surgery in which distended gall bladder is anastomosed to a long loop of jejunum (40 cm) to relieve jaundice. To prevent food particles entering into the gall bladder, enteroenterostomy is done (Fig. 25.61).
   - Most of the patients develop duodenal obstruction caused by the growth in the postoperative period. Hence, a palliative GJ is done at the same time.
   - In the absence of duodenal obstruction, if surgeon thinks that patient may live longer, beyond 6 months, GJ is indicated (for a possible duodenal obstruction occurring later).

II. Nonsurgical treatment

Very elderly patients (age criteria not clear) who are not fit candidates for surgery and patients who have metastasis can be treated by palliative stenting. However, results of a surgical bypass is superior to stenting. Also, the stent needs to be changed frequently.

OTHER CAUSES OF OBSTRUCTIVE JAUNDICE

STRICTURE OF THE CBD

- 80% of strictures occur following surgery on the biliary tree. They are called postoperative strictures. 20% are due to inflammatory pathology.
- It gives rise to slowly progressive, painless jaundice.
- Strictures account for 1–2% cases of obstructive jaundice.

Causes

1. Postoperative post-traumatic (Fig. 25.70)

- **Difficult cholecystectomy:** When the gall bladder is fibrosed, densely stuck to the right hepatic duct or to the common bile duct or as in early cholecystectomy due to oedema around Calot's triangle, injury can occur to the right hepatic duct or to the CBD or CHD resulting in stricture.

Bismuth classification of postoperative stricture (Fig. 25.71)

Type I: Low common bile duct; stump > 2 cm
Type II: Middle common hepatic duct, stump < 2 cm
Type III: Hilar—confluence of right and left duct intact
Type IV: Right and left ducts separated
Type V: Involvement of the intrahepatic ducts

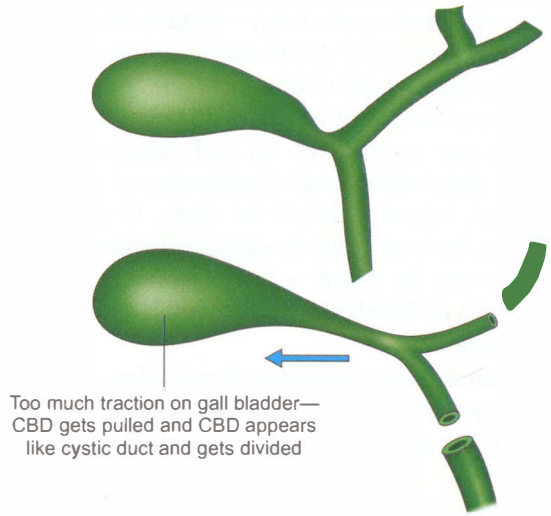


Fig. 25.70: Laparoscopy bile duct injury

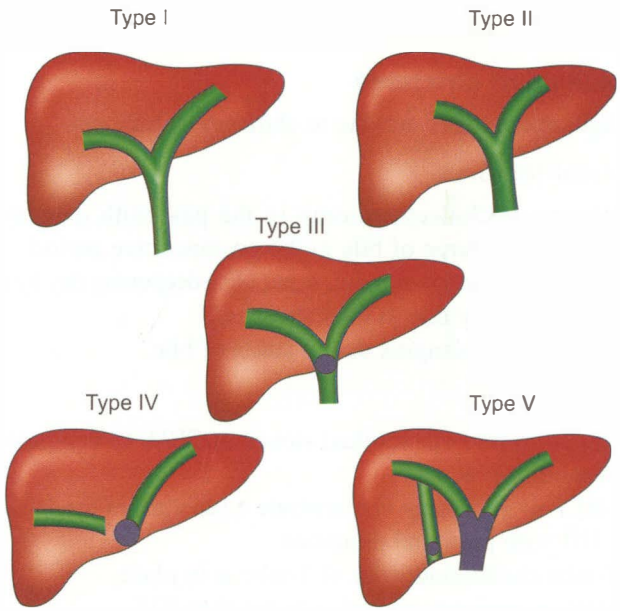


Fig. 25.71: Classification of benign biliary stricture—Bismuth classification (*see* text)

2. Post-inflammatory

Post-inflammatory strictures follow recurrent attacks of cholangitis due to:

- Stones in the CBD or CHD
- Parasites like *Ascaris lumbricoides* in the biliary tree or **Asiatic cholangiohepatitis** produced by Chinese liver fluke infestation (*Clonorchis sinensis*) (page 591).

- Primary sclerosing cholangitis wherein the cause is not known.

PEARLS OF WISDOM

Difficult cholecystectomy, dangerous cholecystectomy and faulty dissection are the important factors for postoperative bile duct strictures.

- **Dangerous cholecystectomy:** Sudden bleeding from cystic artery can occur due to traction on the gall bladder or due to lack of gentleness in ligating cystic artery. Sudden application of an artery forceps to control the bleeding may injure CBD. In such situations, packing the area, good suction and visualisation of the bleeding artery and ligation should be done. If the bleeding continues, the hepatic artery can be compressed between the finger and the thumb in the lesser omentum through the foramen of Winslow. This is called **Hogarth-Pringle manoeuvre**.
- **Dissection at fault:** Ignorance of anomalies such as short cystic duct or too much traction on the gall bladder distorts CBD and predisposes to injury (Figs 25.72 and 25.73).
- It is the duty of the surgeon to show his assistants the Y junction which is formed by the cystic duct, common hepatic duct above and common bile duct below before dividing any structures in this area.

3. Malignant strictures

Malignant strictures are due to cholangiocarcinoma.

Clinical features

- History of cholecystectomy in the past with or without profuse discharge of bile in the postoperative period.
- A slowly progressive, painless jaundice deepening day by day.
- Hepatomegaly due to back pressure.
- Recurrent cholangitis due to stasis of bile.

Investigations

- USG—to rule out residual stones in CBD, to demonstrate intrahepatic dilatation.
- ERCP or PTC may demonstrate a stricture in the CBD or CHD with proximal dilatation.
- T-tube cholangiography, if T-tube is in place.
- MRC is noninvasive and is better than PTC.

Treatment

- If it is due to laparoscopic clipping without transection of the CBD, it is better to re-explore and remove the clips and a T-tube or an endoscopic stent can be placed in the CBD (Fig. 25.74).
- Late cases can be managed by choledochojejunostomy or hepaticojejunostomy by anastomosing a loop of jejunum to the dilated portion above the stricture. However, the general condition of the patient should be improved before surgery (Figs 25.75 to 25.79).

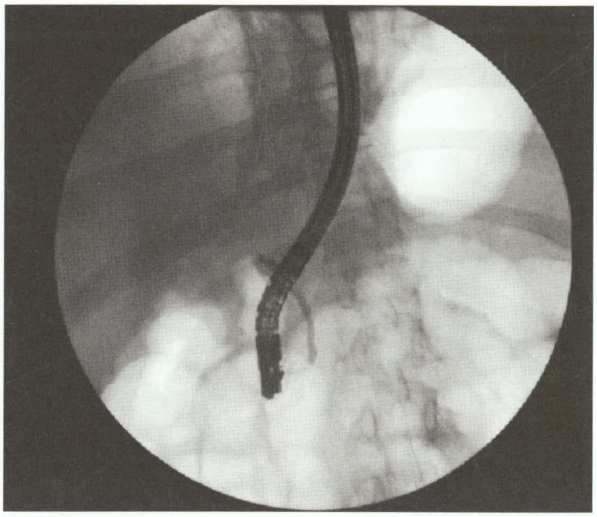


Fig. 25.72: ERCP showing complete transection of bile duct—type IV injury

Fig. 25.73: CBD clipping—here, it was not transected. It was explored, clip removed and endoscopic stent was inserted

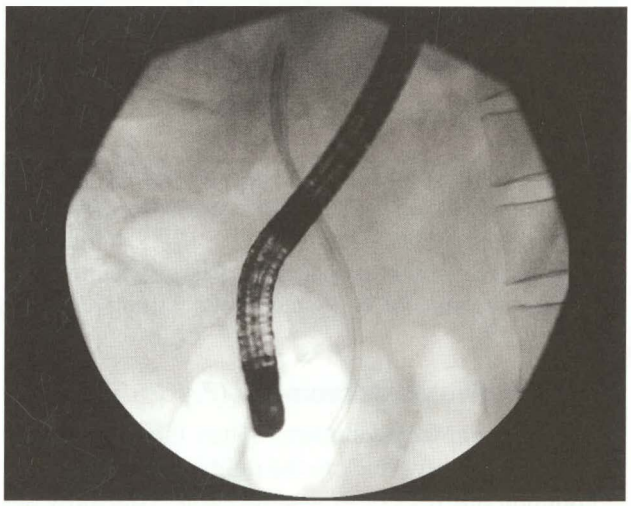


Fig. 25.74: Same patient as in Fig. 25.73. You can see the stent in the common hepatic duct. It was removed after 6 months

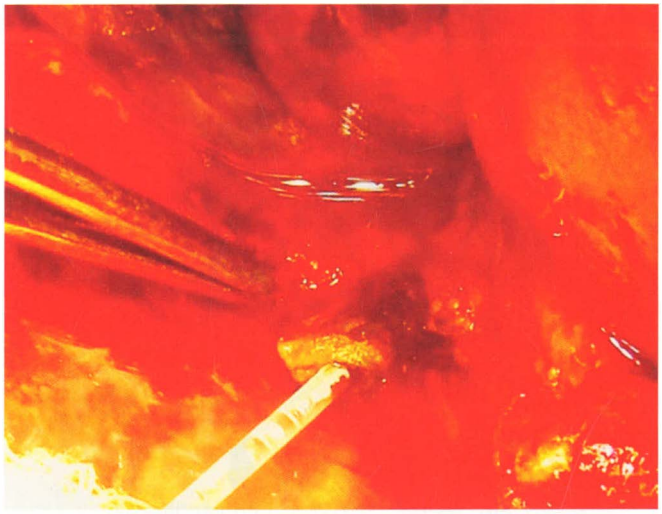


Fig. 25.75: Cut CHD—took one hour to reach this area, it is trimmed for biliary enteric anastomosis

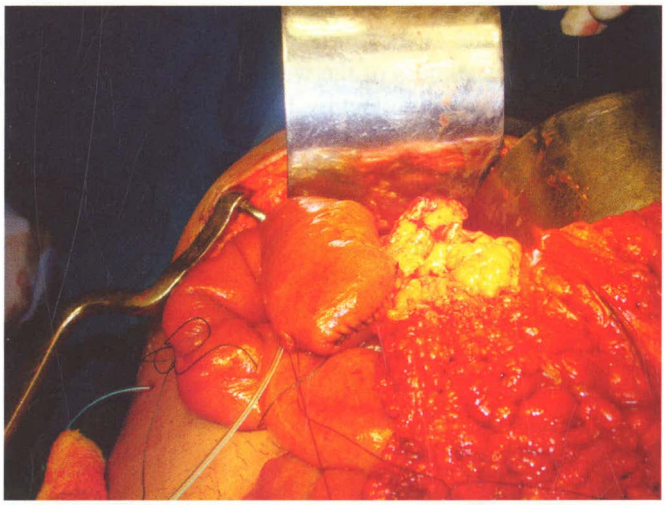


Fig. 25.76: Roux jejunal loop is prepared for hepaticojejunostomy

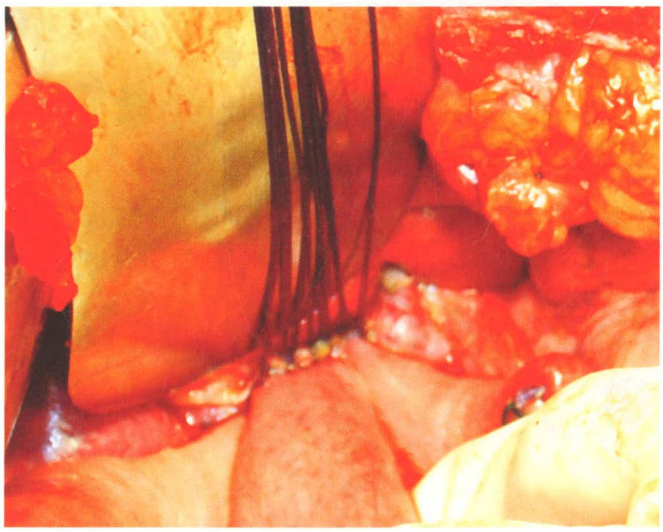
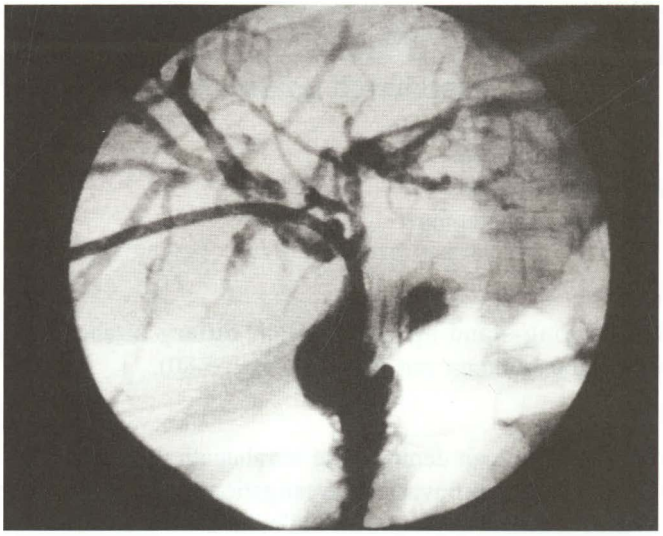
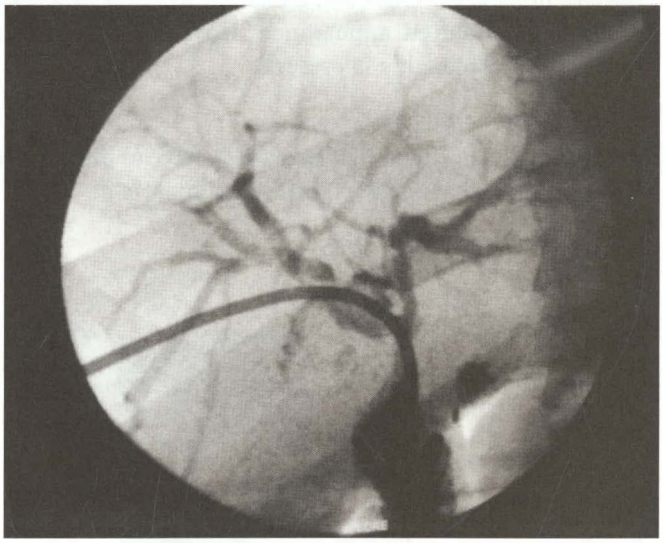


Fig. 25.77: Hepaticojejunostomy was done in another patient with type IV injury





Figs 25.78 and 25.79: PTBD catheter was passed from above across strictured hepaticojejunostomy and dilatations were done in high strictures

PEARLS OF WISDOM

Prevention is better than treatment. Try to prevent bile duct injuries during laparoscopic cholecystectomy.

SCLEROSING CHOLANGITIS

It is characterised by development of multiple strictures and dilatation of CBD with features of fibrous thickening of CBD.

Types

- **Primary**
  No cause is found. However, it can be associated with following conditions (Key Box 25.18).
- **Secondary**
  It is due to stones or injuries.

KEY BOX 25.18

ASSOCIATED CONDITIONS

- Ulcerative colitis
- Crohn's disease
- Graves' disease
- Sjögren's syndrome

Complications

Due to long-standing obstruction, biliary cirrhosis and cholangiocarcinoma can develop (Fig. 25.80).

Diagnosis

- **Ultrasound** can demonstrate intrahepatic dilatation.
- **MRCP** is a noninvasive investigation which can demonstrate multiple strictures and dilatation.
- **ERCP** is the investigation of choice which can demonstrate the strictures in the CBD and dilatation which is described as having a **beaded** appearance. However, the risk of suppurative cholangitis is present.

Treatment: It is difficult

Stenting is the choice although stents may have to be replaced or changed if blockage occurs or if infection sets in.

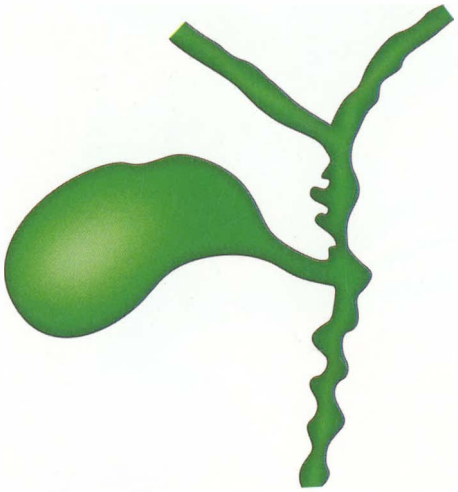


Fig. 25.80: Sclerosing cholangitis

CHOLEDOCHAL CYST

- It is a congenital cyst occurring in the CBD due to partial or complete weakness of the wall of the CBD.
- Majority of cases manifest by 1–2 years of age.

Classification (Fig. 25.81 and Key Box 25.19)

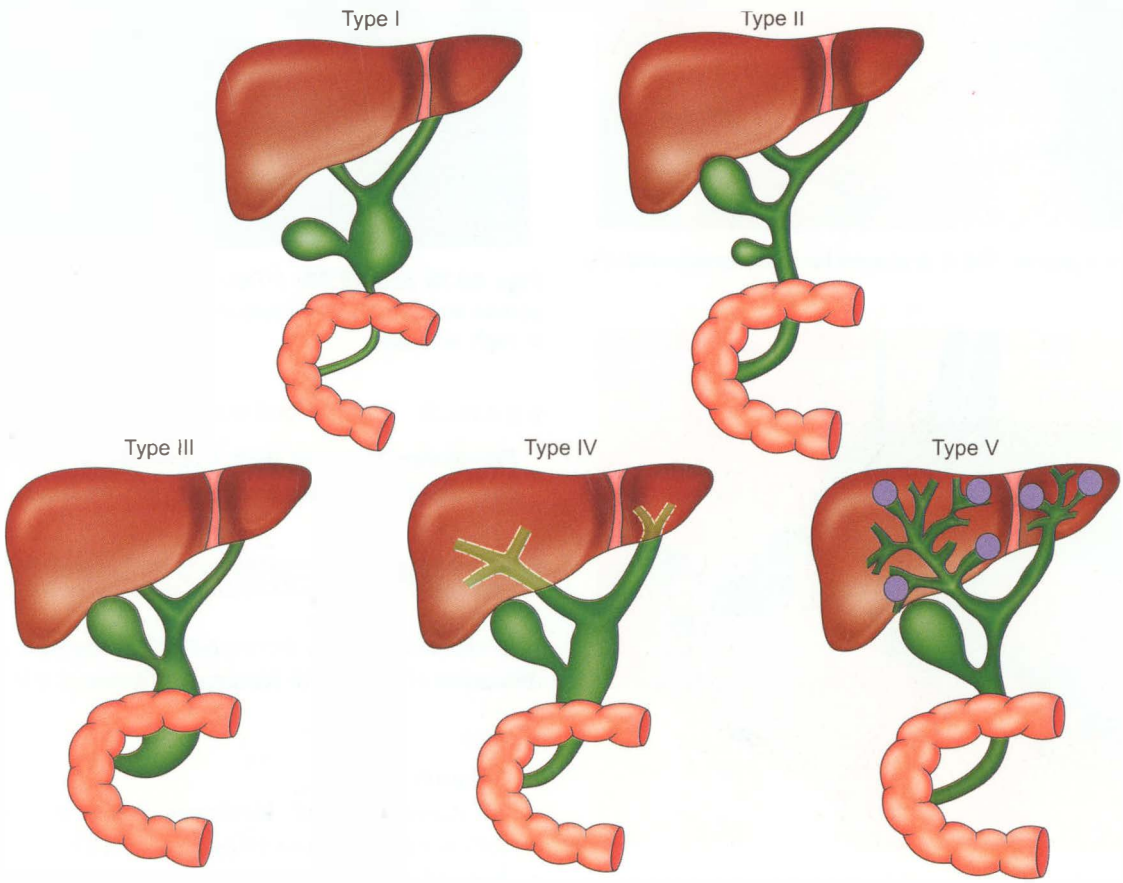


Fig. 25.81: Types of choledochal cysts

KEY BOX 25.19

TODANI CLASSIFICATION

Types

Type I: Fusiform dilatation of CBD—commonest
Type II: Lateral saccular diverticulum of the CBD
Type III: Dilatation of intraduodenal segment of CBD (choledochocoele)
Type IV: Dilatation of CBD + intrahepatic biliary dilatation.
Type V: Multiple intrahepatic cysts—Caroli's disease

Recent advances: Type VI—cystic dilatation of cystic duct

Clinical features

- Age: Majority of cases manifest in children within 1–2 years of age. It can also present in adults.
- More common in **females** 4:1.
- **Abdominal distension** can be due to a large cyst. The cyst can be palpated per abdomen in the right hypochondrium.
- Slow progressive jaundice, recurrent attacks with abdominal pain and pyrexia.

Investigations

1. **USG** will confirm presence of abnormal cyst. It is usually unilocular cyst.
2. **MRC:** It will define the relation between lower end of the bile duct and pancreatic duct to know basic anatomy (Figs 25.82 and 25.83).
3. CT is also useful to know intrahepatic and extrahepatic dilatation.
4. **ERCP** may be done but it will not give any more information than MRC.

Treatment

- **This anomaly is premalignant.** Change to carcinoma is a well-recognised complication and it carries poor prognosis. **Hence, excision of the cyst and reconstruction is the treatment of choice** (Figs 25.84 to 25.87).

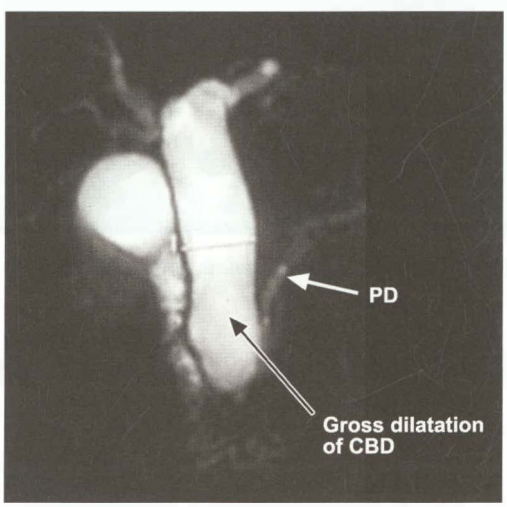


Fig. 25.82: MRC showing choledochal cyst

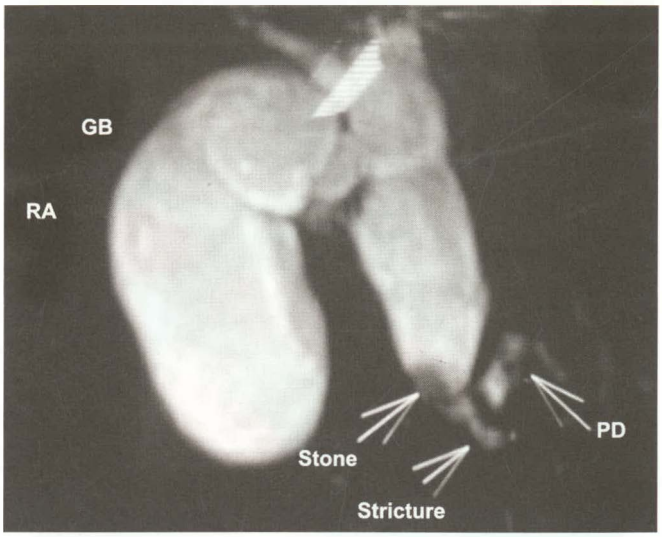


Fig. 25.83: Choledochal cyst—MR cholangiogram showing gross dilatation of the common bile duct and common hepatic duct

- **Type I:** Excision of the cyst followed by Roux-en-Y hepaticojejunostomy.
- **Type II:** Excision of the diverticulum with suturing of CBD.
- **Type III:** Endoscopic sphincterotomy is adequate (choledochocoele).
- **Type IV:** They are difficult to treat. Due to recurrent cholangitis, if total excision is not possible due to adhesions between the cyst and portal vein, **posterior wall of the cyst can be left behind**, after removal of mucosa. This is described as **Lilly's technique**.

Type V will be described later.

Complications

1. **Recurrent cholangitis** with high-grade fever, resulting in biliary cirrhosis.

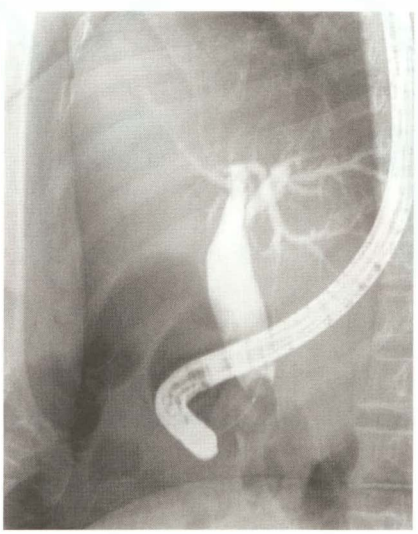


Fig. 25.84: ERCP showing choledochal cyst—type III treated by sphincterotomy

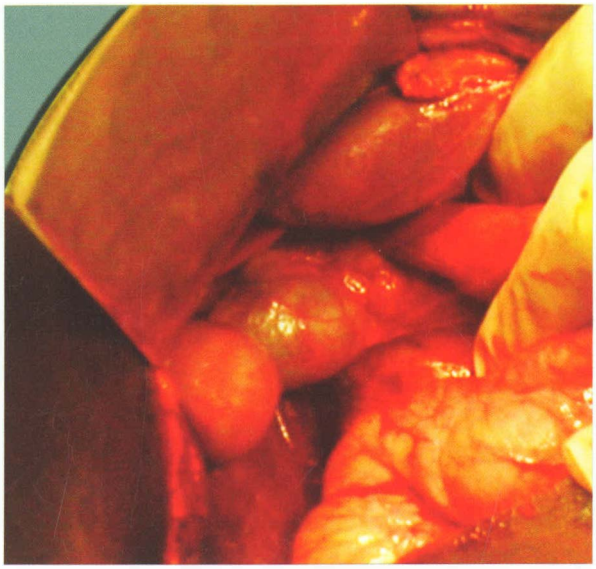


Fig. 25.85: Choledochal cyst at surgery. It was excised completely and hepaticojejunostomy was done (*Courtesy:* Dr Ramesh Rajan, Associate Professor, Surgical Gastroenterology, Trivandrum Medical College, Kerala)

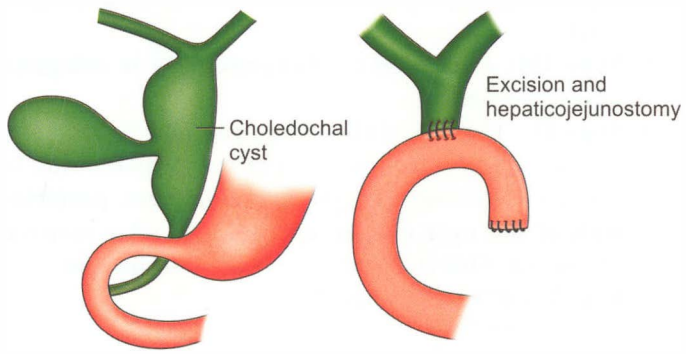


Fig. 25.86: Type I cyst

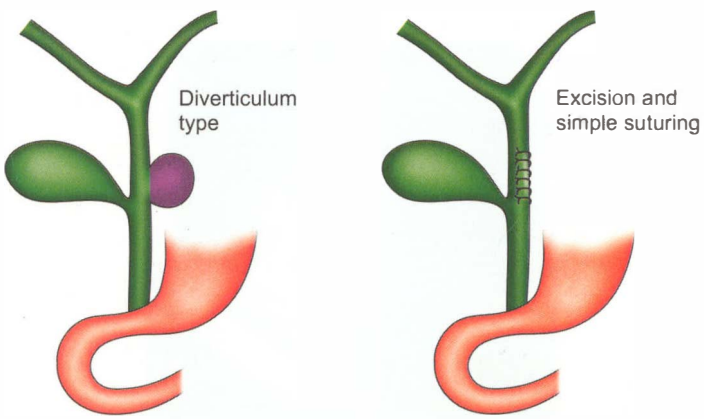


Fig. 25.87: Type II—excision and suturing

2. **Rupture of the cyst** resulting in biliary peritonitis.
3. **CBD stones**
4. **Carcinoma in the cyst** (25–30% of cases). It is a cholangiocarcinoma common in types I and V.

CAROLI'S DISEASE

- It is a hereditary condition wherein there is dilatation o intrahepatic ducts with stenotic segments in between.
- Multiple, irregular saccular dilatations are characteristic.
- It is also an example of ***type V choledochal cyst.***
  Two types:
  1. **Simple:** Presents later with abdominal pain and sepsis.
  2. **Periportal:** It occurs in childhood, presents as recurren cholangitis.
- Diagnosed by ultrasound and CT scan. MRI and ERCP are other investigations (Fig. 25.88).
- Associated lesions are given in Key Box 25.20.

Complications

- Cholangitis: It occurs due to constant obstruction
- Stones: Obstruction and stasis precipitate stone formation
- Biliary cirrhosis
- Cholangiocarcinoma

Treatment

- Hepatectomy (Fig. 25.89)
- Liver transplantation

KEY BOX 25.20

ASSOCIATED LESIONS

- Congenital hepatic fibrosis
- Medullary sponge kidney
- Polycystic liver

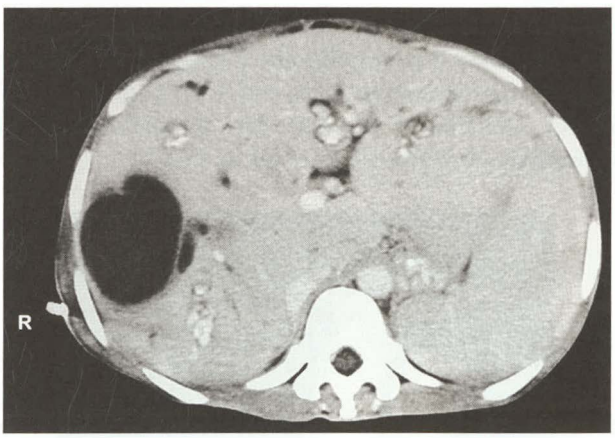


Fig. 25.88: Caroli's disease showing large cyst and intrahepatic dilatation

CHRONIC PANCREATITIS

Definition

Diffuse inflammatory process of pancreas involving head, body and the tail resulting in permanent structural and functional damage to the pancreas.

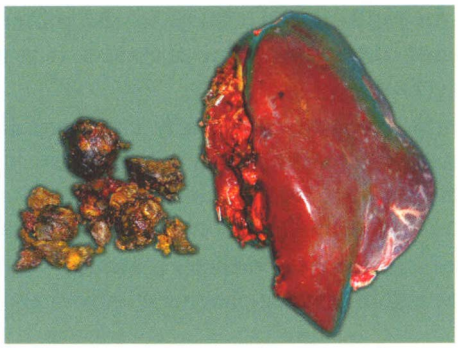


Fig. 25.89: Left lobectomy with extraction of stone—Caroli's disease (*Courtesy:* Dr Nagaraj Palankar, Consultant, Manipal Hospital, Bangalore)

Causes

1. **Alcohol:** High alcohol consumption is the most frequent cause. Alcohol stimulates pancreatic secretion rich in protein. This forms plugs in the pancreatic duct and results in stasis of secretion and stone formation. Alcohol also causes spasm of sphincter of Oddi.
2. **Idiopathic:** It is common in Kerala, and was thought to be due to **consumption of tapioca** (Fig. 25.90). It is also called Kerala pancreatitis or **tropical pancreatitis**. It is now thought to be due to malnutrition. It is also called **fibrocalculous pancreatic diabetes**.
3. **Hereditary pancreatitis:** It is a genetic disorder transmitted as a Mendelian-dominant trait.
4. **Cystic fibrosis:** Generalised dysfunction of exocrine glands cause secretions to precipitate in the lumen.
5. **Hyperparathyroidism** favours precipitation of calcium intraductally. It can also activate pancreatic enzymes.
6. **Autoimmune pancreatitis:** Diffuse enlargement of pancreas and narrowing of pancreatic duct is seen. Auto-antibodies may be present. IgG4 is increased.

Pathology

There is destruction of pancreas by ductal sclerosis, ductal strictures, glandular fibrosis and calcification, both intraductal and parenchymal (Fig. 25.91 and Key Box 25.21).

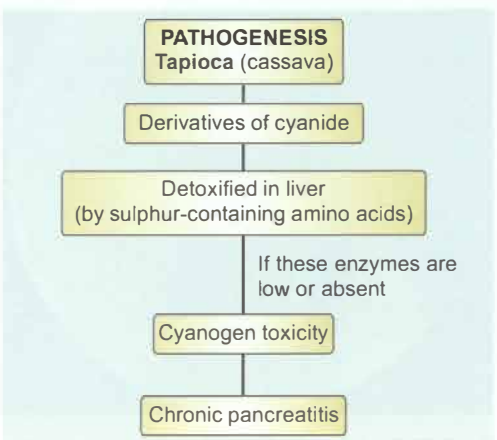


Fig. 25.90: Aetiopathogenesis of cassava pancreatitis

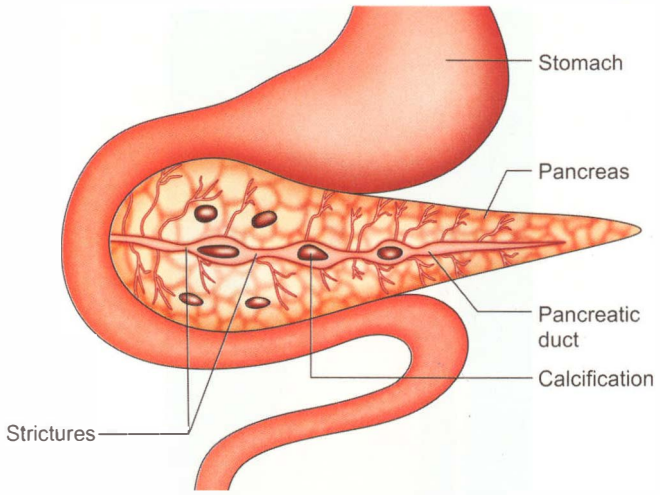


Fig. 25.91: Chronic pancreatitis showing pathological changes

KEY BOX 25.21

CHRONIC PANCREATITIS
IDIOPATHIC TYPE

- Common in **warm** climates (Kerala)
- Common in **young age**
- High incidence of **diabetes**
- High incidence of **stones** in the duct
- Increased chances of **parenchymal calcification**
- Increased chances of **pancreatic cancer**

Clinical features (you can remember as MOPED)

- **M**alabsorption occurs due to damage to exocrine glands resulting in steatorrhoea—10 to 15 stools per day, bulky, frothy, rich in fat, foul-smelling. Malabsorption indicates late disease and results in weight loss. Creatorrhoea refers to excessive loss of protein (Figs 25.92 and 25.93).
- **O**bstructive jaundice can occur due to oedema of the head of pancreas. Later, fibrous constriction of CBD due to fibrotic indurated mass in the head region can cause jaundice by compressing the CBD.
- **P**ain abdomen—upper abdominal pain radiating to the back in the region of L_1 and L_2 due to retroperitoneal inflammation (Key Box 25.22). Pain may be severe, sometimes radiating to both right and left sides. **The pain**

KEY BOX 25.22

PAIN IN CHRONIC PANCREATITIS

- Parenchymal hypertension
- Perineural inflammation
- Ductal hypertension
- Pseudocyst formation
- Stenosis of bile duct

Figs 25.92 and 25.93: This 21-year-old girl was weighing 32 kg in 2006 when she underwent longitudinal pancreatico-jejunostomy (LPJ) for failed medical treatment including stents. After 3 years, she has put on 22 kg weight

Fig. 25.94: Plain X-ray abdomen showing extensive calcification

is due to multiple strictures in the pancreatic duc increasing the intraductal pressure. It is relieved o stooping forward.

- **E**xploratory laparotomy—many cases are diagnosed a laparotomy where irregularity and hardness involving th entire pancreas are seen. Exploration is done for evaluatio of obstructive jaundice or for chronic abdominal pain.
- **D**iabetes—incidence of diabetes is about 10–20%. It shoul be suspected in diabetic patients with pain abdomen.

Investigations

- **Plain X-ray** abdomen can demonstrate stones in the pancreatic duct or parenchymal calcification (Fig. 25.94).
- **USG** can detect the stones, stricture, dilatation and associated cysts.
- **ERCP** (Fig. 25.95)
  - Ductal distension, ductal stricture
  - Dilated pancreatic duct (diameter of the normal duct is 4–6 mm)
  - Demonstration of stones—appear as regular filling defect.
- **CT scan:** It can reveal ductal anatomy, head mass, size and configuration of pancreas (Figs 25.96 and 25.97).

Complications

- Obstructive jaundice due to a mass lesion in the head region
- Carcinoma of pancreas
- Pseudocysts
- Steroids in autoimmune pancreatitis

Treatment

I. Conservative

- Pain relief by analgesics, epidural analgesia, or splanchnic nerve block. Slow release opioid skin patches are useful.
- Supplement **pancreatic enzyme**—diet should be low in fat and **vitamin D** supplements should be given.

Fig. 25.95: ERCP showing dilated pancreatic duct—'chain of lakes' appearance

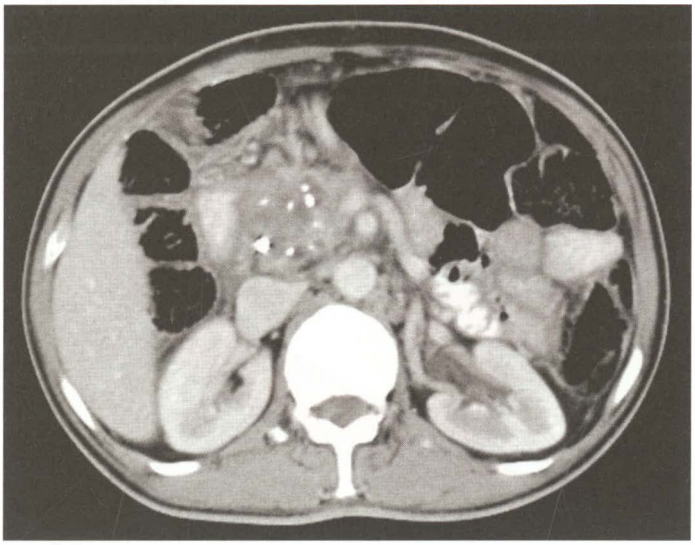


Fig. 25.96: CT scan showing chronic pancreatitis with head mass. This case should be treated with **head-coring** operation

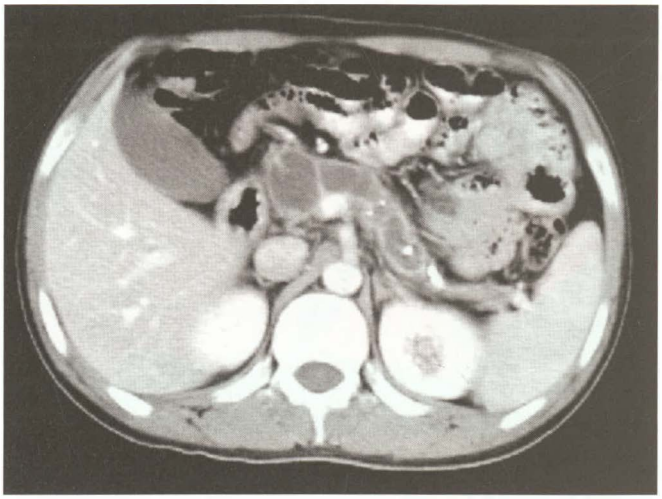


Fig. 25.97: CT shows dilated pancreatic duct of 10 mm in size with stones. ERCP is not required in such patients. This patient underwent LPJ

- Pancreatic enzymes not only help in treating exocrine insufficiency but they also denature CCK releasing peptide, thereby diminishing the release of CCK. Thus, pain is decreased. Six capsules of **pancreatin** are given.
- Control diabetes, stop alcohol consumption and tobacco smoking, antioxidants may help.

II. Surgery: Indications

1. Unrelieved pain
2. Suspicion of carcinoma
3. Complications
   - Ascites
   - Cysts
   - Abscesses
   - GI bleeding—left-sided, portal hypertension, pseudoaneurysm
   - Obstructive jaundice
   - Duodenal obstruction

Types of surgery (Figs 25.98 to 25.107)

1. Chronic pancreatitis involving tail of pancreas
   - Distal pancreatectomy with removal of spleen.
2. **Diffuse chronic pancreatitis with dilated (large duct) pancreatic duct**
   - Duct is laid open widely, strictures are cut open, stones are removed and it is anastomosed to a loop of jejunum—longitudinal pancreaticojejunostomy—**Puestow's operation** (Roux-en-Y jejunal segment). The duct should be at least 8 mm in diameter. Sutures hold very nicely because of fibrosis of pancreas. Pain relief is obtained in about 80% of the cases. Pancreatic fistula is a complication of this surgery. Majority of the fistulae close spontaneously.
   - This is a bypass procedure which preserve endocrine and exocrine functions.

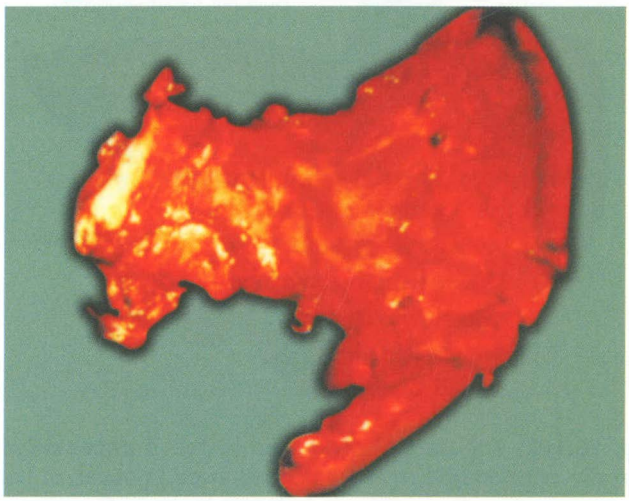


Fig. 25.98: Distal pancreatectomy. Chronic pancreatitis confined to tail of pancreas

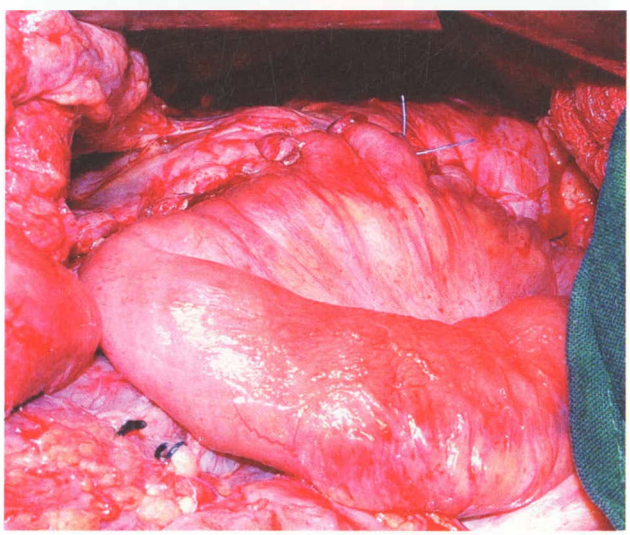


Fig. 25.99: Pancreaticojejunal anastomosis is being done

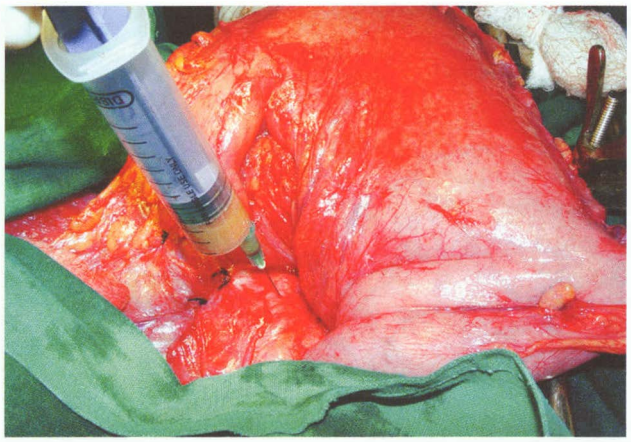


Fig. 25.100: Dilated duct is being aspirated to identify the duct followed by laying it open

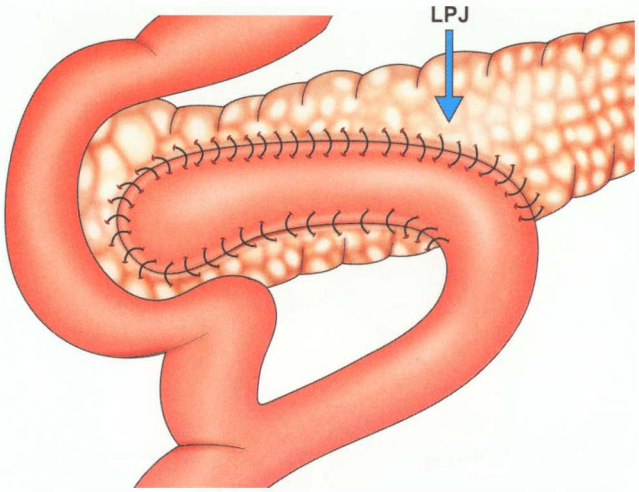


Fig. 25.101: Diagrammatic representation of LPJ

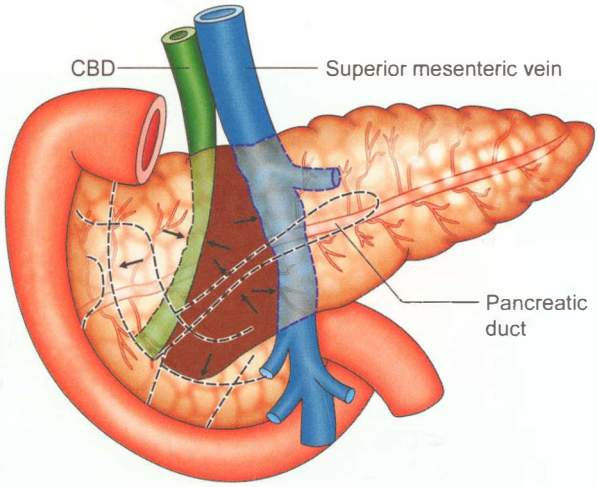


Fig. 25.102: The dark shaded area is called **pacemaker of chronic pancreatitis.** It should be removed in cases of head-coring operation. It is a triangle between lower CBD, pancreatic duct and superior mesenteric vessels

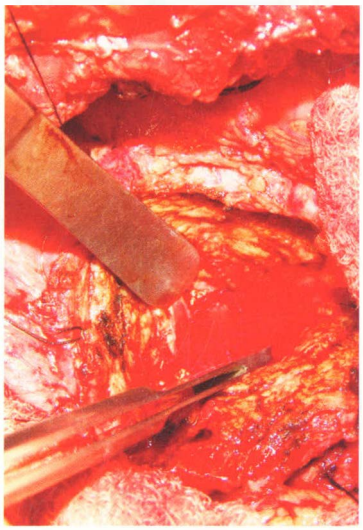


Fig. 25.103: Head-coring is completed. You can watch thin rim of pancreatic tissue all around

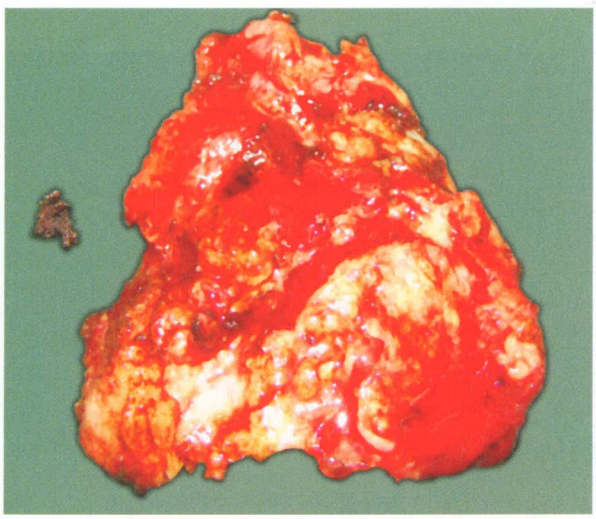


Fig. 25.104: Specimen of the portion of the head which was excised—Frey's procedure (*Courtesy:* Dr Girish MS, MCh, Surgical Gastroenterologist, KMC, Manipal, 2007–2008)

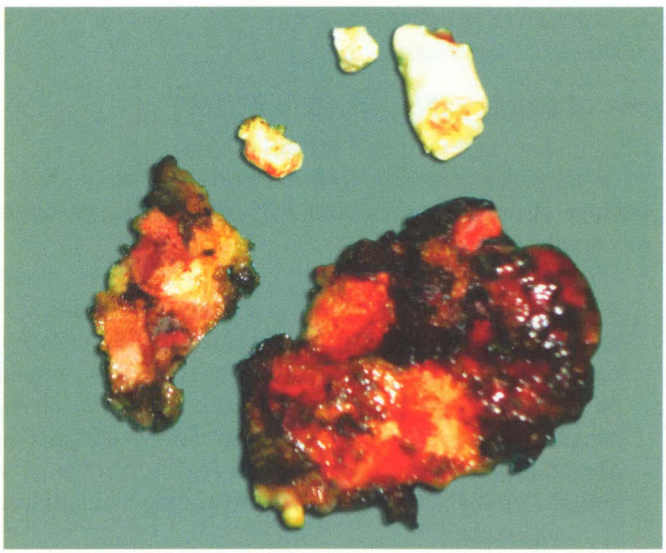


Fig. 25.105: Specimen of the portion of the head along with multiple stones

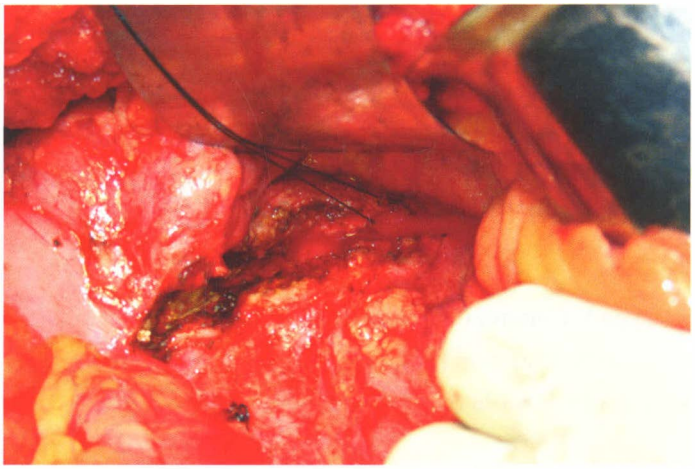


Fig. 25.106: Head-coring gives rise to bleeding which can be controlled with cautery (Key Box 25.23). Pain relief is the main aim of surgery of chronic pancreatitis and a new carcinoma developing. Both these problems are dealt with in this operation

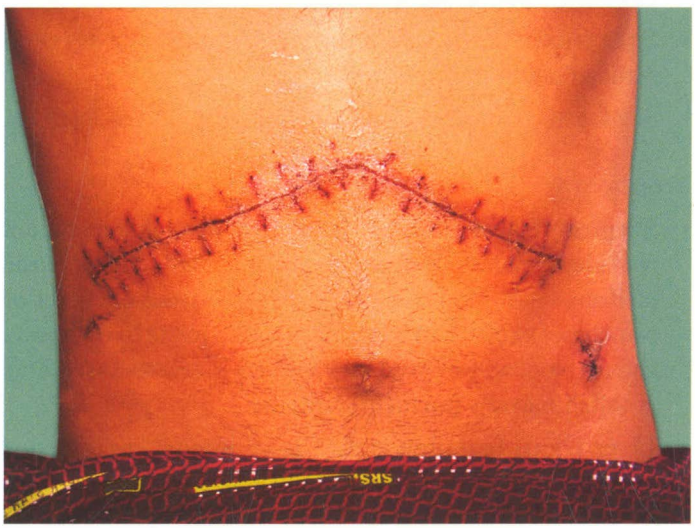


Fig. 25.107: Bilateral subcostal roof top incision gives adequate exposure to all pancreatic surgery

3. **Chronic pancreatitis with a head mass**
   In this situation, doubt arises whether it is malignancy or not. Even trucut biopsy and frozen section are not foolproof. Hence, pancreaticoduodenectomy is advised, provided experience of the surgeon is good and the mortality rate is less than 5%.
4. **Chronic pancreatitis with bile duct obstruction**
   - If malignancy is ruled out, a bypass procedure is the treatment of choice.
     - Choledochojejunostomy is the ideal treatment.
     - Pancreaticoduodenal resection (Whipple) can also be done (as mentioned above).
5. **Chronic pancreatitis with duodenal obstruction**
   Here also, resection of the head mass or gastrojejunostomy is the treatment of choice.
6. **Chronic pancreatitis with ascites:** Treatment of choice is Puestow's operation (stenting may also relieve ascites).
7. **Resection:** A duodenum-preserving pancreatic head resection is called ***Hans Beger's Procedure.*** Head-coring procedure is called ***Frey procedure*** (Figs 25.103 to 25.106 and Key Box 25.23).

KEY BOX 25.23

HEAD-CORING—FREY PROCEDURE

- **Pathologic key** to recurrent pancreatitis is localised to pancreatic head. It is called **pacemaker**.
- It is located in the **crucial triangle** (*see* Fig. 25.102) which has to be excised.
- Pain and recurrent pancreatitis will decrease after Frey procedure.
- Surgery can be done with minimum blood loss.
- Try **avoiding injury to lower CBD**—if necessary, CBD can be opened and a sound can be introduced.

CHOLANGIOCARCINOMA—BILE DUCT CARCINOMA

- Patients having **following diseases** have increased risk of developing cholangiocarcinoma (Key Box 25.24).

KEY BOX 25.24

RISK FACTORS

Cholangitis—primary sclerosing
Colitis—ulcerative
Clonorchiasis
Choledochal cyst
Caroli's disease
Chemical carcinogens like thorium, nitrosamines, diosmin
Observe **6 Cs**

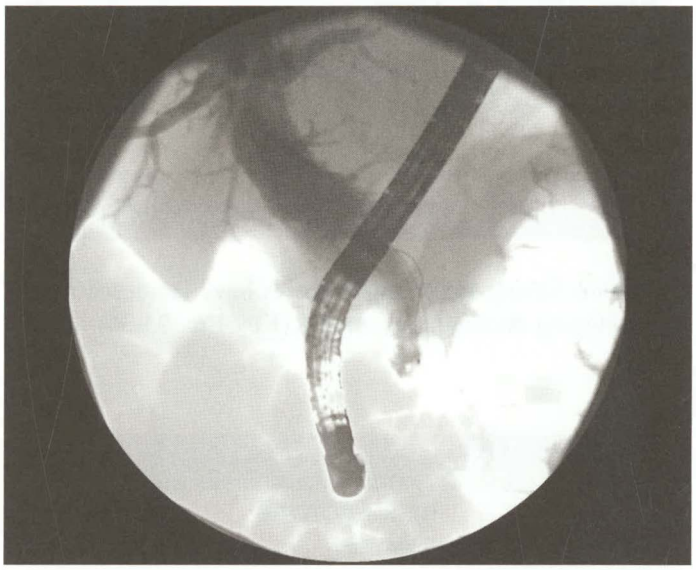
- It is an uncommon cause of obstructive jaundice.
- Elderly males > 60 years are commonly affected.
- **Obstructive jaundice** is the presenting feature.
- Tender hepatomegaly is present due to congestion and may be due to cholangitis.
- **Gall bladder is not palpable** in cases of cholangiocarcinoma unlike in periampullary carcinomas.
- Cholangiocarcinoma at the bifurcation of the hepatic duct is called **Klatskin's tumour.**

Types

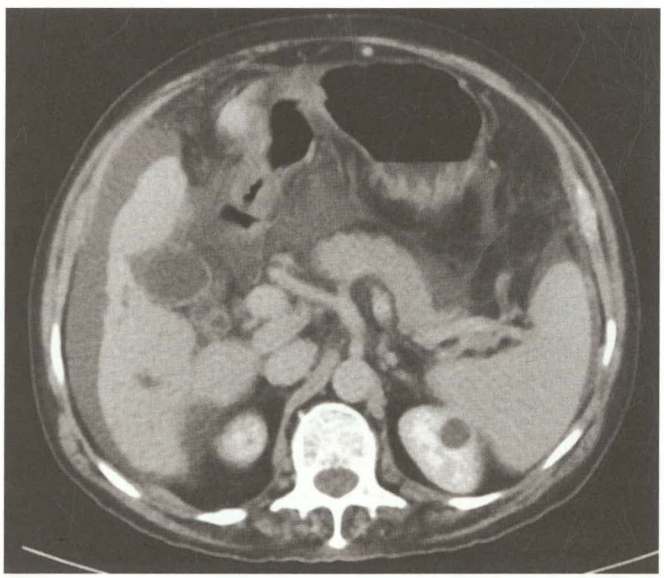
1. Perihilar is the most common, Klatskin's tumour comes under this.
2. Intrahepatic (10%)
3. Distal (CHD-CBD) 25%

Management

- Abdominal ultrasound, CT scan, ERCP will help in localising the site of obstruction (Figs 25.108 and 25.109). ERCP and stenting the CBD is indicated in inoperable cases. However, MRI is better.
- Treatment is difficult because most of the lesions are high up in the hilar region infiltrating the liver, portal vein, etc. Resection and hepaticojejunostomy or endoscopic stent placement are the treatment modalities available.
- Resection involves liver for hilar growth and for lower end growth.



Figs 25.108: ERCP showing cholangiocarcinoma



Figs 25.109: CT scan showing extensive cholagiocarcinoma with ascites

CONGENITAL BILIARY ATRESIA

Aetiopathogenesis

A disease of unknown aetiology, though rare, is fatal. Vira aetiology and defective embryogenesis have been blamed fo the development of biliary atresia.

Types (Fig. 25.110)

- Type I: Common bile duct is involved
- Type II: Common hepatic duct is involved
- Type III: Atresia of right and left hepatic ducts.

Clinical features

- It presents as jaundice at birth or in the neonatal period.
- Due to absence of bile in the gut, meconium is not bile-stained. Hence, stools are pale.
- Gradually, due to backpressure, liver enlarges.
- Enlargement of the spleen may follow if there is development of portal hypertension.
- Steatorrhoea, pruritus and clubbing are the other features.

Treatment

- **Surgical drainage of bile** is the only available treatment provided patent bile duct or radicle is seen. Anastomosis of Roux-en-Y loop of jejunum to the dilated bile duct or sometimes excision of bile duct tissue up to the liver capsule should be done followed by Roux-en-Y anastomosis. This is called **Kasai's portoenterostomy**.
- **Liver transplantation** is the choice when there is atresia of the intrahepatic duct.

Complications

1. Recurrent cholangitis giving rise to hepatic fibrosis
2. Biliary cirrhosis and portal hypertension

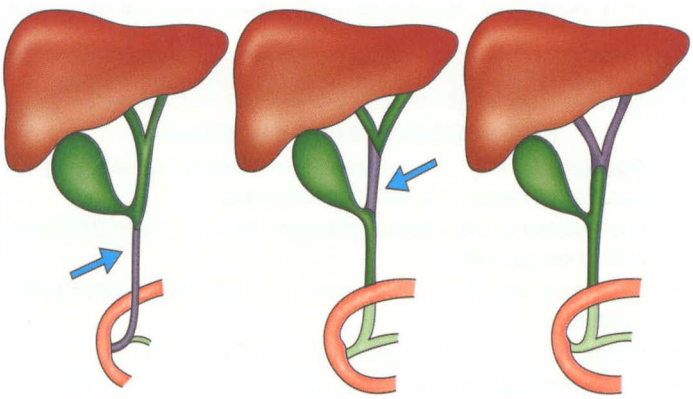


Fig. 25.110: Congenital biliary atresia—types I, II and III

CARCINOMA OF THE GALL BLADDER

Incidence: It is common in North Eastern India. Occurs in elderly patients in their sixties/seventies.

Aetiology

- **Gall stones**
  80–90% of gall bladder cancers are associated with gall stones. Calcification of gall bladder is associated with carcinoma gall bladder.
- **Chemicals**
  High incidence of gall bladder and biliary cancer is noted in people who work in rubber industries.
- **Gall bladder polyp** (Fig. 25.111)
- **Dietary**
  Adulterated mustard oil for cooking is found to precipitate carcinoma gall bladder.

Pathology

- 85% of cases. It is usually adenocarcinoma. Undifferentiated carcinoma and squamous cell carcinoma may also occur. The tumour is most commonly nodular, infiltrates the entire gall bladder and adjacent tissues.
- 3 types of adenocarcinomas have been identified: nonpapillary adenocarcinoma, papillary adenocarcinoma and mucinous adenocarcinoma. Majority are nonpapillary adenocarcinoma.

Clinical features

- Significant weight loss, jaundice and mass in the right upper quadrant are common presentations. Clinically, it is palpable as a hard irregular mass.
- A few cases can present as chronic cholecystitis with a mass or acute cholecystitis. Ultrasonography may diagnose as stones and they are subjected to cholecystectomy. During surgery or postoperatively, the diagnosis gets confirmed.

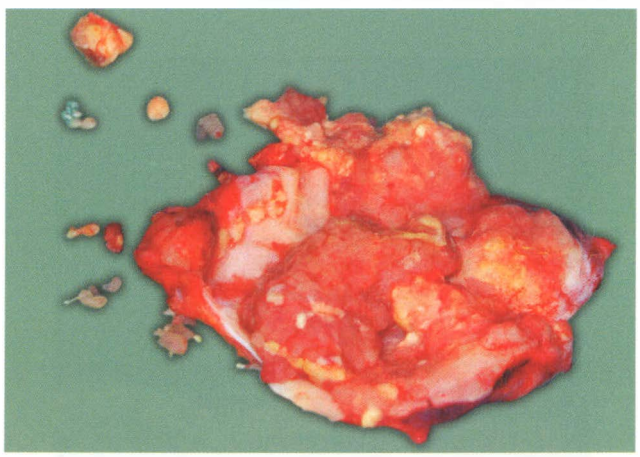


Fig. 25.111: You can see extensive carcinomatous polypoid lesions inside the gall bladder

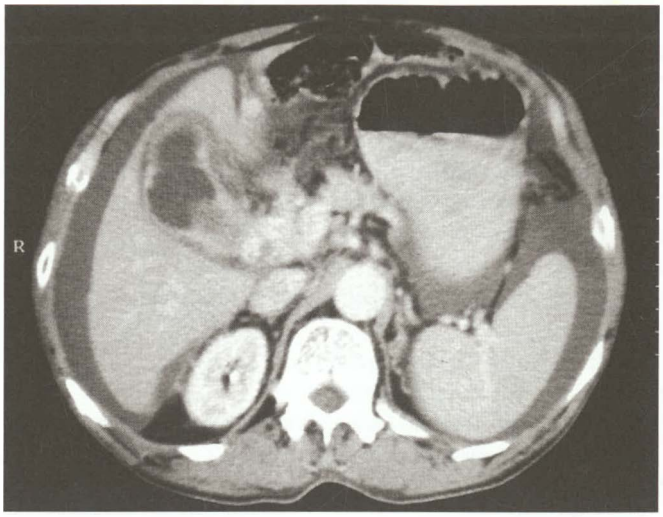


Fig. 25.112: Advanced case of carcinoma gall bladder with ascites and liver infiltration

- Obstructive jaundice, bleeding, ascites are late features.

Investigations (Table 25.2 for staging)

- CA 19–9 is elevated in 80% patients.
- Ultrasonography and endosonography are very useful investigations. Ultrasonography-guided FNAC can be done for histological diagnosis in suspected cases of gall bladder mass.
- CT scan is useful for staging—lymph nodes metastasis in the liver (Fig. 25.112).
- ERCP if there is obstructive jaundice to localise the exact site and nature of obstruction.
- Diagnostic laparoscopy. If peritoneal metastasis is present, it is not worth resecting.
- MRCP can be done. It visualises bile duct better than CT scan.

Treatment

- If gall bladder cancer is found at cholecystectomy and if **mucosa alone is involved**, then cholecystectomy is sufficient.

KEY BOX 25.25

GALL BLADDER CANCER IS THE WORST, WHY?

- Biologically very aggressive cancer (unlike basal cell cancer, carcinoma colon, etc.)
- High incidence of lymphatic spread
- Easily spreads into liver by direct infiltration
- Spreads by blood, neural, intraperitoneal routes
- Intraductal extension into CBD causes obstructive jaundice
- Infiltration into stomach, colon, duodenum and liver can occur because of its location
- Radiation and chemotherapy is rarely of any benefit

Table 25.2 TNM staging of carcinoma gall bladder

TNM staging	
Tumour	**Nodal spread**
Tis – carcinoma *in situ*	N0 – no nodes
T1 – spread to mucosa or muscle layer	N1 – spread to cystic/nodes in portal area
T1a – only mucosal involvement	N2 – spread to parapancreatic/coeliac/superior mesenteric nodes
T3 – spread > 2 cm to liver or 2 or more adjacent organs – CBD, stomach, duodenum, colon, omentum	Metastasis
	M0 – no metastasis
	M1 – distant spread is present
Stage I: T1 N0 M0 (up to muscle)	Simple cholecystectomy
Stage II: T2 N0 M0 (up to serosa)	Extended cholecystectomy
Stage III: T3 N0 – beyond serosa, liver < 2 cm, 1 adjacent organ 1/2/3 N1–hepatoduodenal ligament	Different procedures
Stage IV: T4 N0/1/M0, N2 M1	

- If gall bladder wall is involved, then extended cholecystectomy is done (Key Box 25.25 and Fig. 25.113).
- Radiation has very small benefits.
- Chemotherapy also has been tried. 5-FU, mitomycin C, doxorubicin are the drugs used.

KEY BOX 25.26

EXTENDED CHOLECYSTECTOMY

Cholecystectomy

↓

Open the specimen

↓

Growth penetrating the wall

↓

Segmental wedge excision of liver

↓

Regional lymphadenectomy
(lymph nodes in and around CBD, duodenum, pancreaticoduodenal regions)

The usual blunder committed is doing cholecystectomy for a diagnosis of stone when it is actually a case of carcinoma gall bladder—inadequate surgery results in biliary fistula and hastens death.

Prognosis

In general, five-year survival is very poor (Key Box 25.26). Aggressive surgery and complete clearance give best result (Fig. 25.113).

Thus, we have discussed various causes of obstructive jaundice. Students should be able to differentiate between all these different causes and to offer an appropriate diagnosis in the examination. Each of these short topics can be asked as short notes in the theory paper. Understanding of all these causes needs some concentration power and practice (Key Box 25.26).

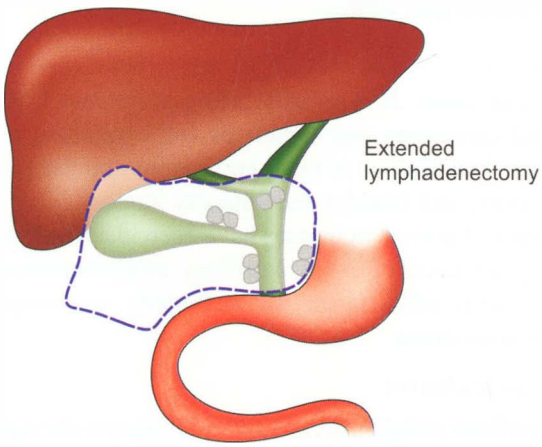


Fig. 25.113: Extended lymphadenectomy

Parasitic infestations causing obstructive jaundice (Table 25.3)

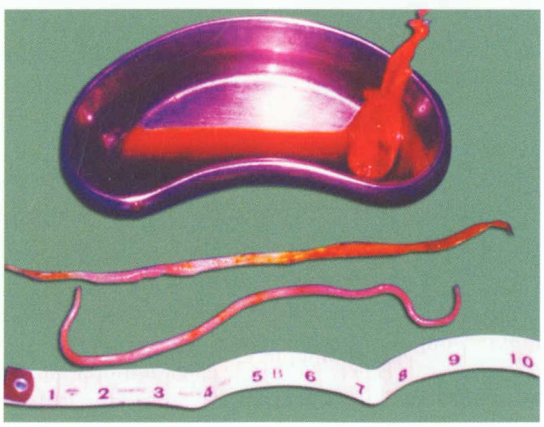


Fig. 25.114: Ascariasis of the biliary duct. (Courtesy: Dr C.G. Narasimhan, Surgeon, Mysore, Karnataka)

Table 25.3 Parasitic infestation of the biliary tract

	Biliary ascariasis	Clonorchiasis	Biliary hydatid
1. Causative organism	*Ascaris lumbricoides* (Fig. 25.114)	*Clonorchis sinensis*	*Echinococcus granulosus*
2. Route of entry	Ampulla of Vater	Ampulla of Vater	Rupture of hydatid cyst of liver into biliary tree
3. Complications	Stricture, suppurative cholangitis, liver abscess, empyema	Biliary pain, stone cholangitis, block, cirrhosis, bile duct carcinogenic	Obstructive jaundice, cholangitis
4. Management	• Ultrasound, CT scan • ERCP—worm can be extracted • Open surgery • Deworming	• Ultrasound, CT scan • Cholecystectomy and T-tube drainage • Choledochojejunostomy for a better drainage	• Ultrasound • ERCP, sphincterotomy, stent • Appropriate surgery

CARCINOMA OF PANCREAS

Introduction

- 70% of the cases occur in the head of the pancreas including periampullary region.
- 30% occur in the body and the tail.
- 70% of cases are adenocarcinoma of duct cell origin.
- It is 4th leading cause of death due to cancer in males, after lung, colon, prostate.

AETIOLOGY

1. **Chronic tropical pancreatitis** and hereditary pancreatitis are associated with pancreatic cancer—such malignancies can be multifocal.
2. **Haemochromatosis** produces extensive calcification of pancreas. It is also a precancerous condition.
3. **Diabetes:** Diabetic patients are 10 times more vulnerable to develop carcinoma of pancreas.
4. Other possible aetiological factors:
   - **Alcohol and smoking:** It is related to tobacco specific nitrosamines (including smokeless tobacco).
   - **Westernisation of diet:** Fatty food, rich in animal proteins can cause pancreatic cancer.
   - **Industrial carcinogens:** B11-naphthylamine, benzidine, gasoline are the possible agents.

PATHOLOGY

- Periampullary refers to carcinoma arising from ampulla of Vater, the duodenal mucosa or the lower end of the common bile duct.
- Microscopically, the types are:
  1. **Mucus** secreting carcinoma of ductal origin.
  2. **Nonmucus** secreting carcinoma of acinar origin.
  3. **Anaplastic** carcinomas are poorly differentiated and tend to arise from the body of the pancreas.
  4. **Cystadenocarcinomas** are rare, slow-growing and tend to attain a large size.

CLINICAL FEATURES

- Periampullary and carcinoma head of pancreas present as obstructive jaundice (discussed already). Discussion here is on carcinoma body and tail of pancreas.
- Microscopically, the tumour shows malignant cells with a variable degree of differentiation, mitotic figures, etc. Not only it invades vessels and lymphatics, but also perineural spread occurs fast which explains backache (Key Box 25.27).

KEY BOX 25.27

CARCINOMA OF PANCREAS—PATHOLOGY

- More than 80% are **ductal adenocarcinomas**.
- **Solid and scirrhous** tumours by nature.
- Plenty of **neoplastic tubular** glands are seen.
- **Desmoplastic** reaction is extensive (feels hard).
- Infiltrates locally—spreads along nerve sheaths, lymphatics and into blood vessels.
- Being retroperitoneal in location, **manifests late, spreads fast and carries poor prognosis**.

Symptoms

- Severe pain radiating to the back in the region of L1 and L2. It is due to infiltration of retroperitoneal nerve plexuses or pancreatic duct obstruction.
- Pain is so severe, often patient requests for narcotic analgesics.
- Gross weight loss in 3–6 months.
- Anorexia, asthenia and generalised weakness.
- Jaundice cannot occur in carcinoma body and tail of pancreas unless there are secondaries in liver or lymph nodes at porta hepatis.
- Symptoms of vomiting suggest duodenal obstruction.

- **Trousseau's sign**[1] (thrombophlebitis migrans)
  - Migrating thrombophlebitis of the legs can occur in visceral malignancies particularly from carcinoma of pancreas, rarely carcinoma stomach, colon, etc.
  - It is supposedly due to sluggish blood flow resulting in thrombus formation.
  - It is superficial and affects the leg veins such as long saphenous vein.

PEARLS OF WISDOM

Sudden development of diabetes mellitus is an early manifestation in 25% of patients.

Signs

- **Anaemia** may be present as in any other malignancy.
- **Jaundice** is not a feature. Left supraclavicular node may be palpable.
- **Per abdomen findings** (Table 25.4): Majority of these cases are advanced, fixed and are felt as a mass in the upper abdomen.

I. Characteristics of a pancreatic mass

- It is situated on the left side involving left hypochondrium, umbilical region and epigastrium.
- It does not move with respiration because it is retroperitoneal.
- It does not fall forwards in knee elbow position.
- Can get above the swelling.
- On percussion, it gives a resonant note because of anterior position of stomach.

II. Features of carcinoma

- It is common in elderly, male patients.
- Hard, irregular, fixed lump.

III. Evidence of metastasis

Secondaries in the liver, ascites, rectovesical deposits

DIFFERENTIAL DIAGNOSIS

1. **Carcinoma stomach** infiltrating the pancreas
   - Such mass may not be mobile. It does not move with respiration because it is fixed to pancreas.
   - These patients will have vomiting first followed by backache at a later date.
2. **Carcinoma transverse colon**
   - Produces constipation and bleeding per rectum. Vertical mobility may be present.
   - Right to left peristalsis may be present.
3. **Para-aortic lymph node mass** may be due to:
   - Intra-abdominal malignancies, lymphoma, testicular tumour, etc.

INVESTIGATIONS

I. Investigations for periampullary carcinoma and carcinoma of head of pancreas (discussed before).

II. **Investigations for carcinoma of body and tail**

1. **USG:** First investigation of choice
   - Rules out calculous obstruction
   - Can detect a mass as small as 2 cm
   - Intraoperative ultrasound can be used to take a biopsy.
2. **Contrast-enhanced CT scan** (Figs 25.115 and 25.116)
   - Retroperitoneal invasion
   - Lymph node enlargement
   - Invasion of hepatic artery, superior mesenteric artery, ascites, liver metastasis—unresectable.
   - Infiltration of portal vein which makes it inoperable
   - In more than 70% of cases pancreatic duct is dilated and in more than 60% of cases bile duct is dilated.
   - When both these ducts are dilated, it is most likely pancreatic cancer.
   - CT-guided aspiration biopsy is indicated for lesions to start neoadjuvant chemotherapy/RT (infiltrative lesions).

Table 25.4 Differences between pancreatic mass and stomach mass

Carcinoma body of the pancreas	Carcinoma stomach
1. Pain radiating to the back is the presenting feature	Vomiting, dyspepsia is present. Pain is not a feature
2. Haematemesis is not a feature	Haematemesis is a common feature
3. The mass does not move with respiration	Usually moves with respiration
4. Mobility is not a feature	Mobility is present
5. Knee elbow test—the mass does not fall forwards	Stomach mass falls forward (intraperitoneal), unless it is fixed
6. Retroperitoneal organ	Intraperitoneal
7. Percussion: Resonant note is obtained because of stomach/intestine anterior to it	Impaired note because of growth in the stomach

[1]**Trousseau's sign** is also found in Buerger's disease

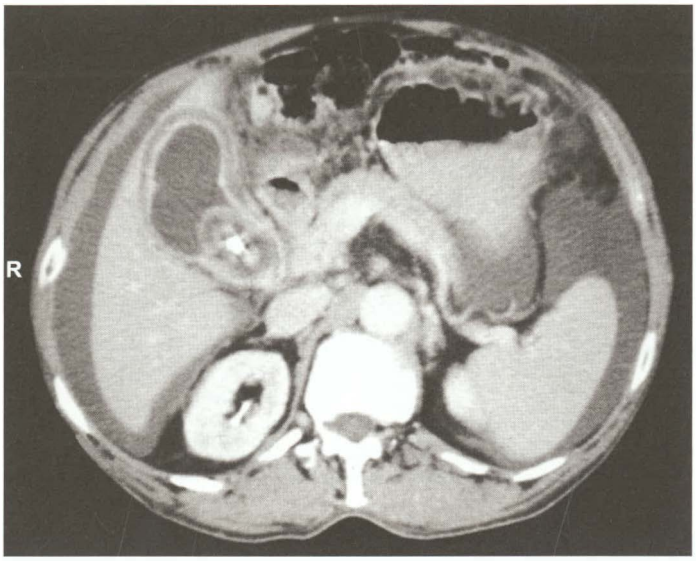


Fig. 25.115: Carcinoma body of the pancreas with para-aortic nodes and ascites

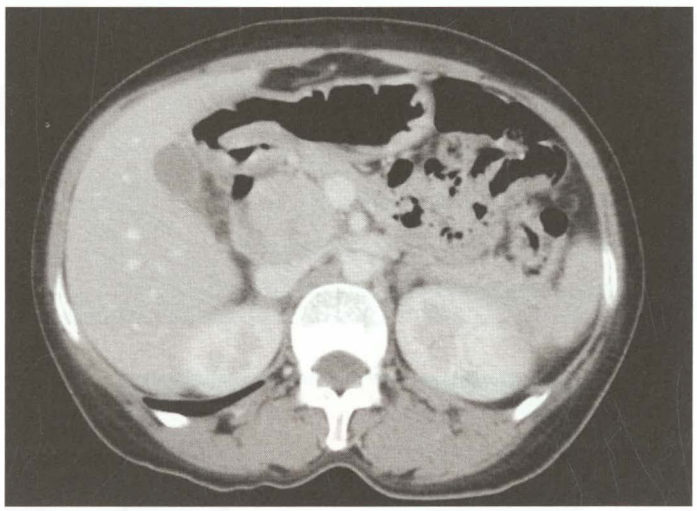


Fig. 25.116: Large mass in the head and body of pancreas

3. ERCP is indicated in the absence of a mass. Findings can be stenosis/obstruction.

4. **Carbohydrate antigen CA 19–9:** These are glycoproteins which are elaborated by malignancies. ***It is a tumour marker of pancreas to monitor carcinoma pancreas.*** Increased in 75% of patients with carcinoma pancreas and 10% of the patients in benign disease of pancreas, liver and bile ducts. Hence, it is not a diagnostic investigation. However, very high values suggest malignancy even when histology is inconclusive.

TREATMENT

I. Periampullary carcinoma—Whipple's operation

II. Carcinoma of body and tail

A. If the tumour is very small and diagnosed very early, they are ideally treated with **total pancreatectomy with removal of involved lymph nodes.**

B. Many cases are diagnosed late. They are inoperable either due to fixity to portal vein or due to metastasis. Hence, there is no role for curative surgery. For the confirmation of diagnosis, transabdominal USG-guided fine needle aspiration cytology can be done. Surgery is not indicated in such cases; only palliative treatment can be offered. Prognosis is very poor.

- **Palliative radiotherapy:** 4000–6000 cGy units can be given. Response rate is 5–10%. It reduces size of the tumour and **some pain relief is obtained** (Table 25.5).
- Palliative treatment is an important aspect of carcinoma pancreas even though such patients succumb to the disease within 3 to 6 months. Palliation is mainly to relieve pain.

TRUE CYSTIC PANCREATIC NEOPLASM (CPN)

- These are uncommon tumours present predominantly in women (Key Box 25.28).
- Other rare tumours are cystadenocarcinoma, acinar cell cystadenocarcinoma, etc.
- These tumours can be confused with pseudocyst of the pancreas. Hence, they should be differentiated at surgery (Figs 25.117 to 25.120). Otherwise, gross blunders can take place (Table 25.6).
- Serous and mucinous are important. They are discussed in Table 25.7.

Table 25.5 Palliation for carcinoma pancreas—3 chief symptoms

Pain	Jaundice	Duodenal obstruction
• Sustained release oral narcotic drugs—morphine sulfate	• Stenting—recurrent obstruction and cholangitis chances are decreased	• Occurs in 20% patients
• Coeliac plexus nerve block (pain is due to invasion of retroperitoneal nerve trunks)	• Choledochojejunostomy, cholecystojejunostomy (if cystic duct enters CBD well above the growth)	• Posterior dependent GJ is preferable—indicated in cases of symptoms such as vomiting
• Intraoperative coeliac plexus block by injecting 50% alcohol directly into tissues along side of aorta	• Surgical bypass has increased mortality and morbidity initially	
• Palliative RT and gemcitabine—survival improved by 1–2 months		

Table 25.6 Differences between cystic neoplasm and pseudocyst

	Cystic neoplasm	Pseudocyst
1. History of pancreatitis	Absent	Present
2. Imaging	May demonstrate *solid component*	Usually absent
3. ERCP	Mucin *egress* from bulging papilla in cases of IPMTs	No
4. Mucin content of fluid	Present	Absent
5. CEA in fluid	Increased in mucinous cystic neoplasms	Absent
6. CA 19–9	Increased in malignant cystic neoplasms	Not elevated
7. Percutaneous wall biopsy	Epithelial lining can be present	No epithelial lining

Table 25.7 Cystic neoplasms

	Serous	Mucinous
• Gender	• No prediction	• More common in women
• Nature	• Multiple cysts are present	• More often multilocular
• Cut surface	• Microcystic adenomas with cut surface appearance of a sponge	• Macrocystic adenomas with smooth lining with papillary projections
• CT scan	• CT may show cysts with calcification	• Septal on CT scan are characteristic
• Nature of fluid	• Cystic spaces contain serous fluid	• Cystic spaces contain mucus
• Lining epithelium	• Lining is by cuboidal epithelium	• Lining is tall columnar and Goblet cells
• Malignant potential	• No malignant potential	• Most of them turn into cyst adenocarcinoma

KEY BOX 25.28

TRUE CYSTIC PANCREATIC NEOPLASMS (CPNs)

A. Serous Cystadenoma: Benign

- Common in women, located in head of pancreas. They account for 30% of all CPNs. They can be observed if asymptomatic.

B. Mucinous Cystic Neoplasms (MCNs)

- More common in women
- More often found in body and tail
- More incidence than serous (40%)
- Considered premalignant
- Do not communicate with ductal system
- Lesions more than 2 cm need to be resected

C. Intraductal Papillary Mucinous Tumours (IPMTs)

- Slightly more common in males
- Communicates with duct
- Incidence is about 25%
- High malignant potential
- More common in head involving ampulla of Vater
- Treated by Whipple's resection

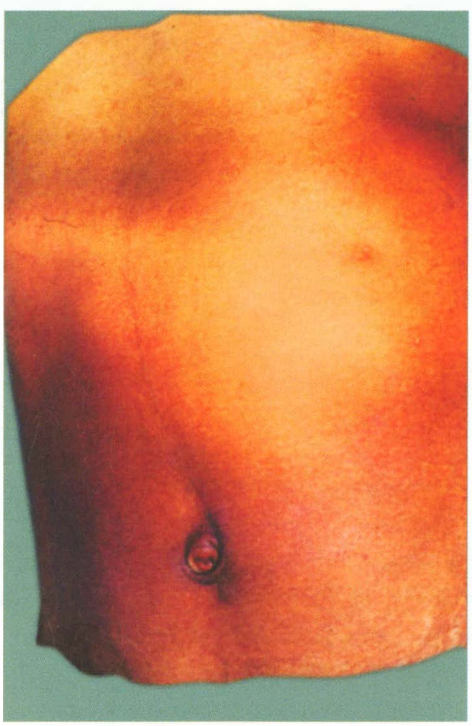


Fig. 25.117: Cystadenoma of the pancreas

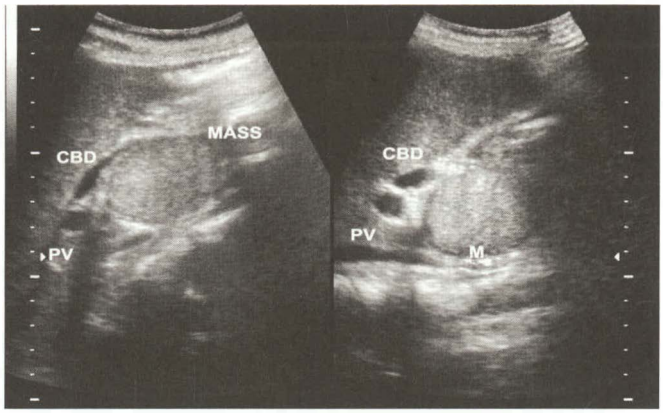


Fig. 25.118: Ultrasound showing cystic mass in the body of pancreas

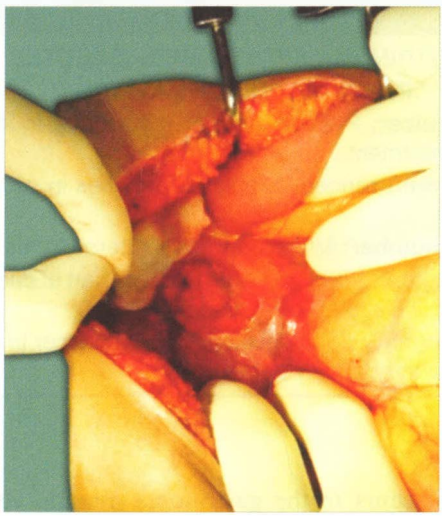


Fig. 25.119: Cystadenoma of the pancreas at surgery

ENDOCRINE TUMOURS OF THE PANCREAS

These tumours are members of APUDOMA arising from APUD (**A**mine **P**recursor **U**ptake **D**ecarboxylation) cells. They are neuroectodermal in origin. Accordingly, following tumours can occur (Table 25.8).

Types

- **Sporadic:** Usually occurs as a single tumour.
- **Familial:** Occur with other adenomas as in multiple endocrine neoplasia syndrome—**MEN TYPE I:** Pituitary, parathyroid, pancreatic adenoma.

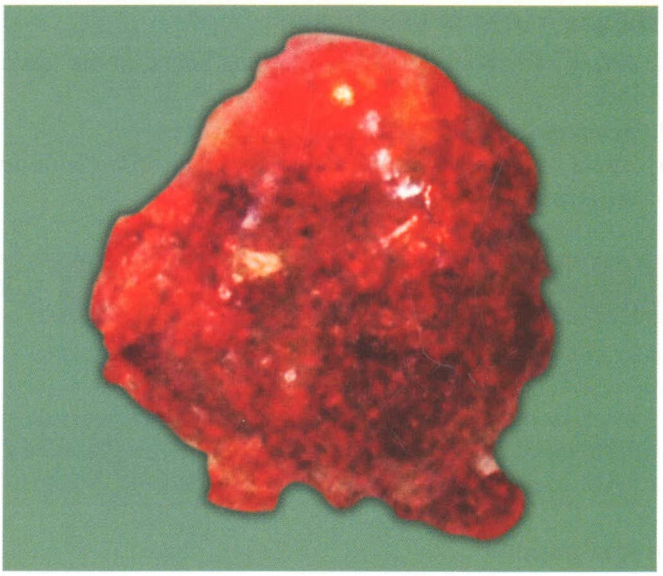


Fig. 25.120: Cystadenoma of the pancreas resected specimen

INSULINOMA (β-CELL TUMOUR)

- Most common functioning islet cell tumour.
- Majority of the insulinomas occur in the tail and body of pancreas and majority of them are benign, one-third are malignant and one-third are multiple.
- It can be a single adenoma, can be due to diffuse hyperplasia or due to carcinoma.
- **Clinical features are that of hypoglycaemia** (Key Box 25.29 for other causes of hypoglycaemia).
- In early stages, it can mimic a duodenal ulcer. Due to hypoglycaemia, hunger pain develops and a tendency to ask for food is present.
- As it becomes severe, giddiness, dizziness, syncopal attacks, blurring of vision and diplopia can occur.
- Late stages—epilepsy, semiconsciousness and coma.

KEY BOX 25.29

PERSISTENT HYPOGLYCAEMIA—CAUSES

- Insulinoma
- Hepatoma
- Hepatocellular damage
- Hypopituitarism
- Addison's disease
- Large mesenchymal tumour

Table 25.8 Summary of islet cells

Islet cells	Active agent	Syndrome	Presentation
Alpha	Glucagon	Glucagonoma	Hyperglycaemia
Beta	Insulin	Insulinoma	Hypoglycaemia
Delta	Somatostatin	Somatostatinoma	—
G	Gastrin	Gastrinoma	Hyperacidity

Whipple's triad of insulinoma

1. Attack of hypoglycaemia in morning hours, in the fasting state.
2. Symptoms are relieved on taking glucose.
3. Blood sugar in the fasting state is less than 45 mg% during the attack.

CLINICAL NOTES

The author remembers a case of recurrent epilepsy getting admitted to the department of neurology. He was found to have hypoglycaemia and was diagnosed as insulinoma only during the 4th admission. A 3 cm benign insulinoma was enucleated from the pancreas. The patient is asymptomatic till date.

Investigations

1. **Serum insulin** levels done by immunoassay method are found to be very high. Proinsulin levels and C-peptide level are also very high.
2. Persistent hypoglycaemia—blood sugar level of less than 50 mg/dl relieved by glucose is suggestive of insulinoma.
3. **USG and CT scan** of abdomen can demonstrate the tumour if it is more than 2 cm (intraoperative ultrasound is a useful test).
4. Selective angiography will demonstrate tumour blush as majority of them are very vascular.

Treatment

- Enucleation is the treatment of choice.
- Resection of the tumour and if necessary distal pancreatectomy can also be done in selected patients.
- Diazoxide is given to suppress insulin release.
- For inoperable islet cell carcinoma, streptozocin is the best chemotherapeutic agents.

Insulinoma—interesting most

- The most common functioning islet cell tumour of pancreas.
- Mostly are single (75%)
- Mostly mutiple in 'MEN' syndromes
- Mostly benign (70%)
- Mostly occur in tail and body of pancreas.
- Mostly present as bizarre behaviour confusing type due to hypoglycaemia.
- Most useful diagnostic test—very high (disproportional) levels of insulin during fasting hypoglycaemia.
- The most important test to localise the tumour before surgery is CT/MRI.
- The most important test to localise at surgery is intraoperative ultrasound.
- Mostly they can be enucleated when occur in the head.
- Most insulinomas are small, less than 2 cm

GASTRINOMA (ZOLLINGER-ELLISON SYNDROME

Two types are recognised

- **Type I (rare):** These have antral G cell hyperplasia, wherei gastrin is stored. Hypergastrinaemia with chronic pepti ulceration is a feature. Pancreas is normal.
- **Type II: May be due to an ulcerogenic,** non β-islet cel tumour or sometimes diffuse hyperplasia of the islet cells Tumour secretes gastrin and is usually found in the tail o the pancreas. 50% of them are malignant.
  - Intractable peptic ulceration, hypergastrinaemia wit massive acid hypersecretion of up to 500 ml/hr can occu
  - Diarrhoea, steatorrhoea and hypokalaemia occur due t acid irritating the small bowel activity (Key Box 25.30)

KEY BOX 25.30

GASTRINOMA SHOULD BE SUSPECTED IN FOLLOWING SITUATIONS

1. **Unusual ulcer:** Peptic ulcer not responding to intensive medical treatment.
2. **Unusual recurrence:** Multiple recurrences in spite of the treatment.
3. **Unusual number:** Multiple ulcers scattered in the GIT.
4. **Unusual sites:** Ulcers present in the 2nd part of duodenum, an ulcer just distal to the ligament of Treitz.
5. **Unusual age:** Sudden development of an ulcer in a young boy or in a very elderly patient.

Site

It commonly occurs in the gastrinoma triangle also called Passaro's triangle. Following three points form **Passaro's triangle.**

1. Junction of the cystic duct with the common bile duct (Fig. 25.121).

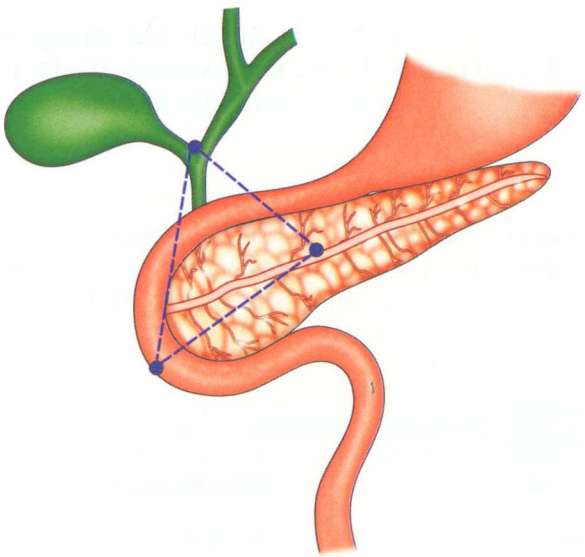


Fig. 25.121: Passaro's triangle of gastrinoma
(*Courtesy:* Dr. Vidushi, Undergraduate student, KMC, Manipal)

2. Junction between head and neck of pancreas.
3. Junction between second and third parts of the duodenum.

Diagnosis

1. **Endoscopy** will show prominent mucosal folds and large amount of acid in the stomach.
2. **Serum gastrin** is increased above normal levels (normal value < 150 ng/dl).
3. **CT scan and arteriography** may localise the tumour.

Treatment

1. **Type I:** Partial gastrectomy to remove G cells bearing area.
2. **Type II:** If the tumour is small, removal of the tumour can be done (enucleation).
3. If the **tumour is large**, omeprazole 20 mg twice a day or somatostatin derivatives have been used to control acidity. Total gastrectomy is the last resort, if gastrinoma is not found.

GLUCAGONOMA

- It arises from **alpha cells** producing glucagon. Around 90% are in the body and tail.
- Clinically, it presents with **extensive necrolytic migratory erythematous rashes.** Rashes occur due to low amino acid levels due to neoglucogenesis brought about by glucagon. Rashes involve legs and perineum.
- Age range is 20–70 years
- This is common in women
- **Mild diabetes**, weight loss, diarrhoea are the other features.
- Glucagon levels are increased in serum diagnostic.
- Localisation is by elective arteriography and CT scan
- **Treatment is enucleation**

Treatment principles of glucagonoma

- Correction of malnutrition by TPN (total parenteral nutrition).
- Treatment with somatostatin analogues
- Enucleation is easy if it is in the head
- Considerable palliation even if subtotal removal is achieved.
- Low dose heparin—before and after surgery—because of high risk of DVT—pulmonary embolism.
- Streptozocin and dacarbazine are the most effective chemotherapeutic agents.

Thus, endocrine tumours of the pancreas are rare but when they occur, they can present with varying clinical manifestations which confuse a clinician. A high index of suspicion is necessary. They may be benign but can be fatal if left untreated.

ACUTE PANCREATITIS

Definition: It is defined as acute nonbacterial inflammatory condition caused by activation, interstitial liberation and autodigestion of pancreas presenting as acute abdominal pain.

- *Acute pancreatitis stings like a scorpion (produces severe pain).*
- *Acute pancreatitis drinks like a fish (produces dehydration).*
- *Acute pancreatitis eats like a wolf (pancreatic necrosis).*
- *Acute pancreatitis burrows like a rodent (produces fistula).*
- *Acute pancreatitis kills like a leopard (life-threatening).*

Marseille's classification of pancreatitis

1. Acute pancreatitis
2. Acute relapsing pancreatitis
   - In both these conditions, pancreas returns back to normal.
3. Chronic pancreatitis
4. Chronic relapsing pancreatitis
   - In both these conditions there is always permanent damage to the pancreas.

AETIOLOGY (Figs 25.122 to 25.124)

1. **Alcohol abuse (40 to 50%):** It is the **major cause of acute pancreatitis** in **our country** and is seen in about 50% of the cases. Alcohol stimulates pancreatic secretions rich in protein, forms protein plugs and results in obstruction to the pancreatic duct. Alcohol stimulates trypsinogen. It causes spasm of sphincter of Oddi. It also has **direct toxic effect on the pancreas**. Tobacco smoking contributes to its effects.
2. **Biliary tract disease:** Stone in the biliary tree (gall stone pancreatitis) is the major cause of acute pancreatitis in the Western world (40%). In our country, it may be responsible for pancreatitis in about 20–30% of patients.

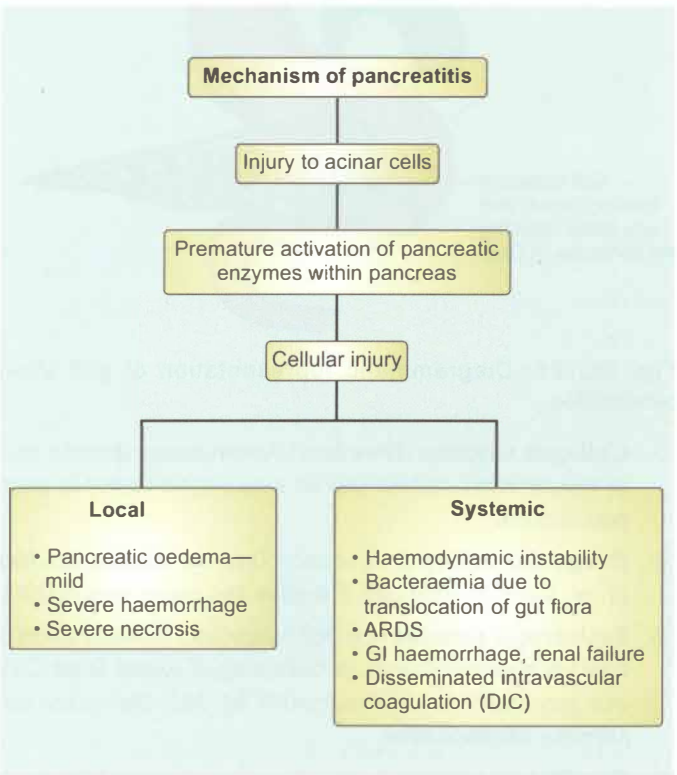


Fig. 25.122: Pathogenesis of tissue damage (*see* text)

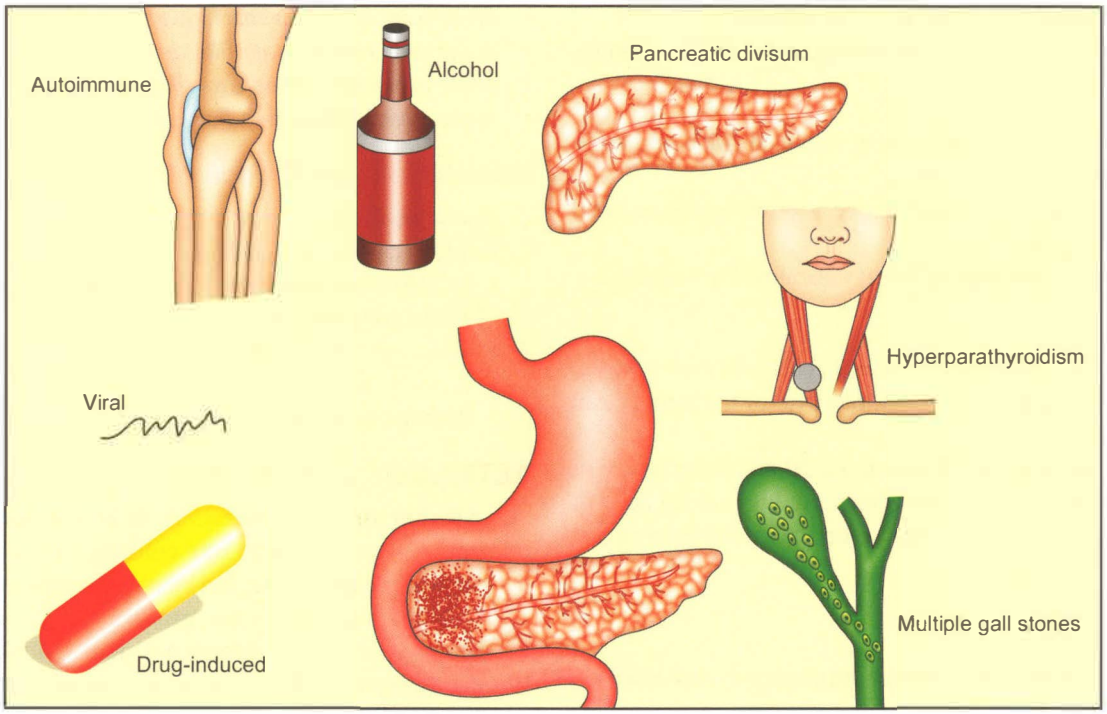


Fig. 25.123: A few causes of acute pancreatitis

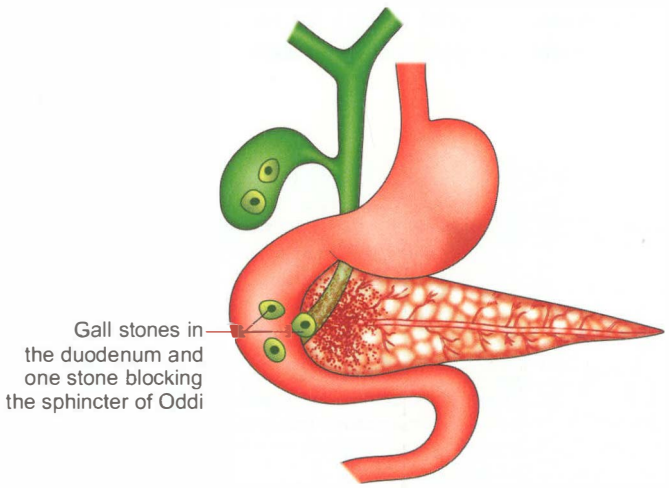


Fig. 25.124: Diagrammatic representation of gall stone pancreatitis

3. **Collagen vascular disorders:** Autoimmune disease such as polyarteritis nodosa can be a causative factor in acute pancreatitis.
4. **Drugs:** Corticosteroids, tetracycline, oestrogens, azathioprine, valproic acid and diuretics can cause pancreatitis.
5. **Endoscopic procedures:** Sphincterotomy, cannulation of CBD or pancreatic duct, or basketing of stones from CBD can precipitate acute pancreatitis by duct disruption and enzyme extravasation.
6. **Familial or genetic** factors have been blamed for acute pancreatitis. Hereditary pancreatitis may be due to mutation of cationic trypsinogen gene. Symptoms begir in early childhood.
7. **Hyperparathyroidism** causing hypercalcaemia may stimulate pancreatic juices and can cause pancreatitis. I also facilitates precipitation of calcium in the ducts.
8. **Hyperlipidaemia** (> 1000 mg/dl of triglycerides) can alsc cause pancreatitis. Dietary control can cure pancreatitis.
9. **Hypothermia** and **hypotension** can cause ischaemia to the pancreas resulting in acute pancreatitis.
10. **Injury to the pancreas** either postoperative or following penetrating injury can result in pancreatitis.
11. **Infection:** Virus such as *Mumps* and *Coxsackie* can cause pancreatic. Scorpion sting can also cause pancreatitis abnormality.
12. **Ductal:** Around 20%. Some of them are due to pancreatic divisum seen during ERCP. The opening of the minor papilla is inadequate for drainage of pancreatic juice.
13. **Postoperative pancreatitis**
    - It follows operations on CBD-open or laparoscopic. More common after T-tube insertions (now it is less).
    - Sphincteroplasty, ERCP, stone extractions
    - Pancreatic biopsy
    - Gastrectomy (distal/total)
    - Cardiac surgery. Here risk factors are perioperative administration of calcium, postoperative hypotension, preoperative renal failure.

14. **Idiopathic:** It is seen in about 15% of cases. Even though, classified as idiopathic (no cause is found), they are found to have sludge/gall stones undetected by ultrasound examination (Key Box 25.31).

KEY BOX 25.31

QUICK REFERENCE TO COMMON CAUSES OF PANCREATITIS

- Pancreatitis in teens—suspect **hereditary or APBDJ**[1]
- Pancreatitis in women—suspect **gall stones**
- Pancreatitis in males—alcoholic pancreatitis
- Pancreatitis with **fleeting joint pains—autoimmune pancreatitis.**
- Pancreatitis with bony lesions/cysts—**hyperparathyroidism**
- Pancreatitis with fever—viral

[1]Abnormal pancreatico-biliary duct junction

Pathogenesis (Fig. 25.125)

- Autodigestion is the final common pathway leading to pancreatitis, a few theories have been explained as possible factors for autogenesis.

1. **Obstruction/secretion:** Recurrent attacks of pancreatitis as in chronic cases can be due to multiple strictures, stasis, resulting in autodigestion.
2. **Common channel theory:** In about 90% of cases, bile duct and pancreatic duct converge into ampulla of Vater. However, only in 10% of cases they have common channel. Reflux of bile into pancreas has been blamed for 'gall stone pancreatitis'. However, strong evidence is lacking.
3. **Duodenal reflux:** Activation of enzymes takes place in the duodenum through action of enterokinase. Hence, it is postulated that duodenal reflux is one of the causes of acute pancreatitis.
4. **Back diffusion:** Back diffusion of pancreatic enzymes through the ductal epithelium is a possible factor, when damaged by alcohol/bile acids, etc.
5. **Systemic sepsis:** Organ failure
   - ARDS, renal failure
   - Endotoxins originate from bacteria.

- Acute pancreatitis is an **autodigestion** following activation of trypsinogen. This is brought about by various agents mentioned above.
- It may also be due to reflux of bile into the pancreas.
- Trypsinogen is converted into trypsin. It acts and stimulates:

1. **Lipase:** Lipase splits the fat into fatty acids and glycerol. Fatty acids combine with calcium to form calcium soap. This is represented as fat necrosis seen in the omentum, subsynovial pockets of knee joint, etc. This also explains hypocalcaemia and tetany seen in acute pancreatitis.
2. **Elastase:** It digests the elastic fibres of the blood vessels resulting in rupture and haemorrhage into the peritoneal cavity.

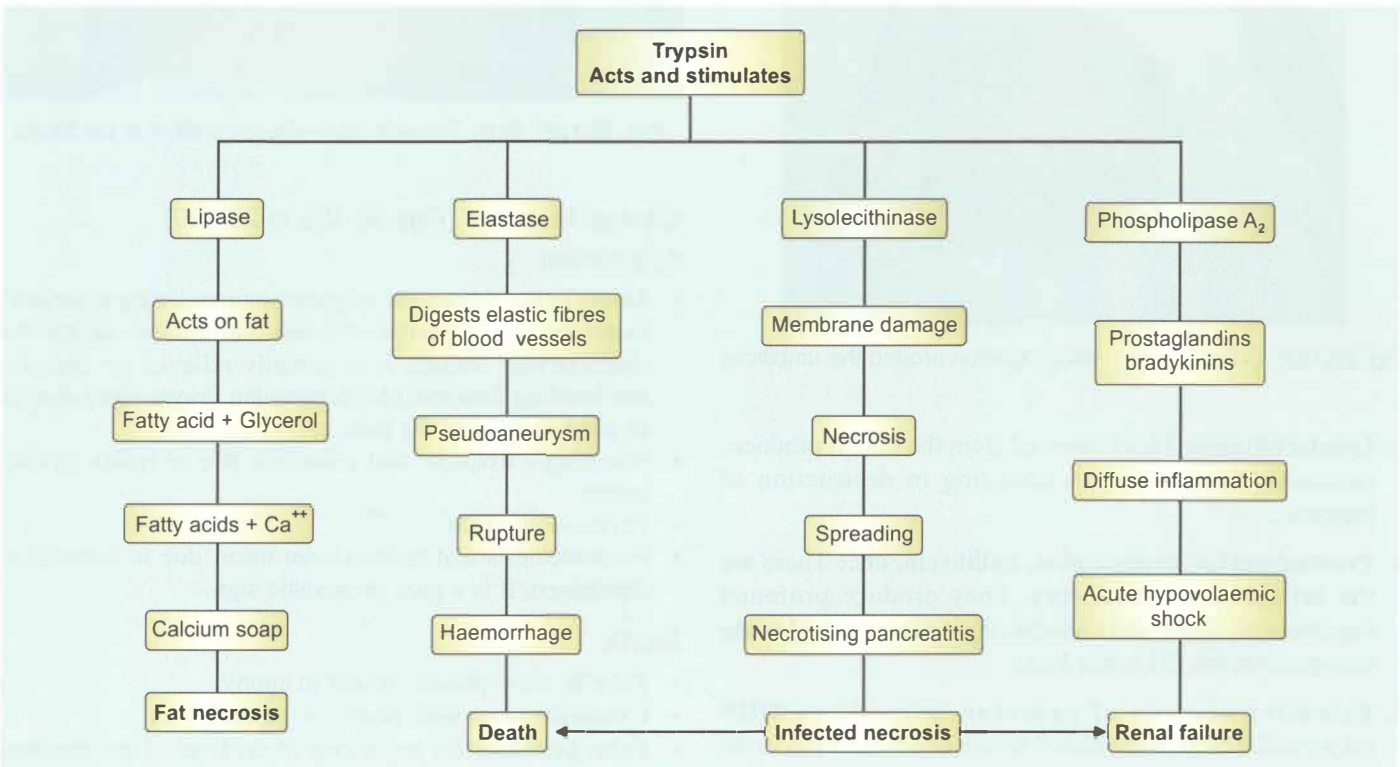


Fig. 25.125: Activation of trypsinogen to trypsin—pathogenesis of pancreatitis

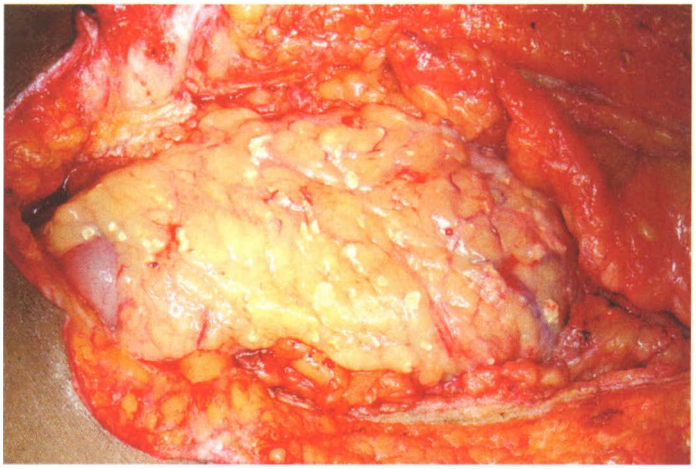


Fig. 25.126: Fat necrosis in the omentum

Fig. 25.128: Cullen's sign—discolouration around the umbilicus

3. **Lysolecithinase:** This is derived from the bile. It produces extensive tissue necrosis resulting in destruction of pancreas.
4. **Prostaglandins, bradykinins, kallikrein, etc.:** These are the inflammatory mediators. They produce profound hypotension, shock and collapse, due to loss of fluid in the retroperitoneum (III space loss).
5. **Extensive necrosis of pancreas:** It produces **MDF** (**m**yocardial **d**epressant **f**actor) which depresses ventricular contraction resulting in cardiac failure. Ultimate result is the development of **multi-organ failure**.

Fig. 25.127: Abdominal distension is due to paralytic ileus in the early stages. It can also be due to pancreatic ascites

Fig. 25.129: Grey Turner's sign—discolouration in the flanks

Clinical features (Figs 25.126 to 25.129)

Symptoms

- Severe upper abdominal epigastric pain radiating to the back increases over a period of hours—illimitable agony is a characteristic feature. It is partially relieved on stooping and bending forwards (Mohammedan Prayer sign). A meal or alcohol triggers the pain.
- Vomiting—frequent and effortless due to reflex pyloro-spasm.
- Fever—low grade
- Haematemesis and melaena can occur due to necrosis of duodenum. It is a poor prognostic sign.

Signs

- **Febrile**, tachypnoeic patient in agony.
- **Cyanosis**—improper perfusion of lungs.
- **Faint jaundice** due to oedema of the head of the pancreas.
- **Features of shock**—feeble pulse, tachycardia, hypotension, cold extremities.

- **Abdominal findings**
  - Tenderness in epigastrium
  - Upper abdominal guarding and rigidity
  - Distension of the abdomen
  - Mass in epigastrium
  - Muscle guarding
  - Abdominal distension due to either accumulation of blood or fluid in the peritoneal cavity or due to paralytic ileus.
- **Cullen's sign** (Fig. 25.128)
  - Bluish ecchymotic discolouration seen around umbilicus (first described for ruptured ectopic pregnancy).
- **Grey Turner's sign** (Fig. 25.129)
  - Bluish discolouration in the flanks
  - Both these signs are due to peri-pancreatic and retroperitoneal haemorrhage and seepage of blood along fascial planes, into the anterior abdominal wall and spread through falciform ligament.
- **Evidence of respiratory signs:** Tachypnoea, dullness, effusion, crepitations, rhonchi (pulmonary oedema, ARDS).

Investigations

1. **Haemogram (CBP)**: Hb% may be low due to haemorrhagic pancreatitis.
   - Total count is raised above 15,000 cells/mm^3 due to inflammation.
2. **Blood for urea, creatinine** to rule out renal failure
3. **Serum amylase** (widely used test) (Key Box 25.32).
   - Normal levels are 40–80 Somogyi units
   - Values around 400 are suggestive and values more than 1000 Somogyi units are diagnostic of acute pancreatitis.
   - It is increased in the first 24–48 hours and returns to normal within 3–4 days (Key Box 25.32).
   - Persistent high level of amylase in acute pancreatitis indicates:
     - Unresolving inflammation
     - Recurrent attacks of pancreatitis
     - Complications—pseudocyst, pancreatic abscess
     - **Serum lipase levels—more specific** but difficult to measure. Lipase is only **secreted by pancreas.**

KEY BOX 25.32

INCREASED AMYLASE LEVELS ARE SEEN IN

- Acute pancreatitis and its complications
- Parotitis
- Afferent loop obstruction
- Spasm of sphincter of Oddi
- Biliary peritonitis—duodenal injuries
- Mesenteric infarction
- Ruptured ectopic gestation

4. **Blood and urine sugar estimation:** Glycosuria is present in almost 100% of patients.

PEARLS OF WISDOM

Lipase levels tend to be higher in alcoholic pancreatitis and amylase levels are higher in gall stone pancreatitis.

5. **Serum calcium levels:** Hypocalcaemia is seen, due to hypoalbuminaemia or fat necrosis.
6. Total proteins are usually low, especially albumin.
7. **Plain X-ray abdomen (erect position)** (Fig. 25.130)
   - **Calcification suggests chronic pancreatitis.**
   - To rule out perforation of peptic ulcer.
   - **Sentinel loop sign**—one dilated jejunal loop of intestine which is seen in the region of pancreas.
   - **Colon 'cut-off' sign** refers to mild distension of transverse colon with collapsed descending colon.
8. **Abdominal ultrasound**—can demonstrate oedematous pancreas, fluid in the abdomen or biliary tract disease.
9. **Contrast enhanced CT scan** of abdomen is done after 3–5 days in patients who fail to respond to conservative treatment. **If CT scan demonstrates infected necrosis, an urgent CT-guided FNAC is done and Gram stain is sent. If Gram stain is positive, it has to be treated urgently** (Key Box 25.33, Figs 25.131 and 25.132).

Indications for CT scan in acute pancreatitis

- Patients with severe pancreatitis
- When the diagnosis is in doubt
- Patients with organ failure
- Patients with sepsis
- Localised complications such as pseudocyst, pseudo-aneurysms.
- Clinical deterioration

Fig. 25.130: Plain X-ray abdomen erect showing dilated jejunal loop due to paralytic ileus—sentinel loop sign

KEY BOX 25.33

BALTHAZAR CT SCAN SCORING SYSTEM OF ACUTE PANCREATITIS

CT Grade		Points
A	Normal	0
B	Oedematous	1
C	Mild extrapancreatic collection	2
D	Severe extrapancreatic collection	3
E	Extensive/multiple extrapancreatic collection	4

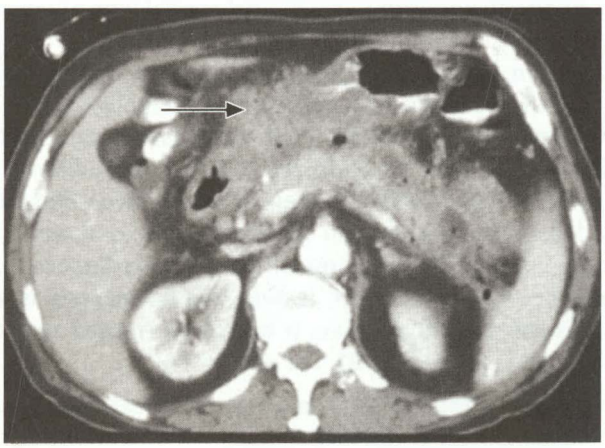


Fig. 25.131: Contrast enhanced CT (CECT) scan is the gold standard investigation in cases of acute pancreatitis. Useful after 7 days of acute pancreatitis. This photograph shows extensive necrosis. CT scan also helps in detecting pseudocysts, abscesses, presence of gall stones or any other unsuspected pathology in the abdomen. Modern thinking is to wait for 4–6 weeks for separation of necrosis before surgical necrosectomy

Management

- Almost always conservative. However, a few scoring systems have been followed (Table 25.9).

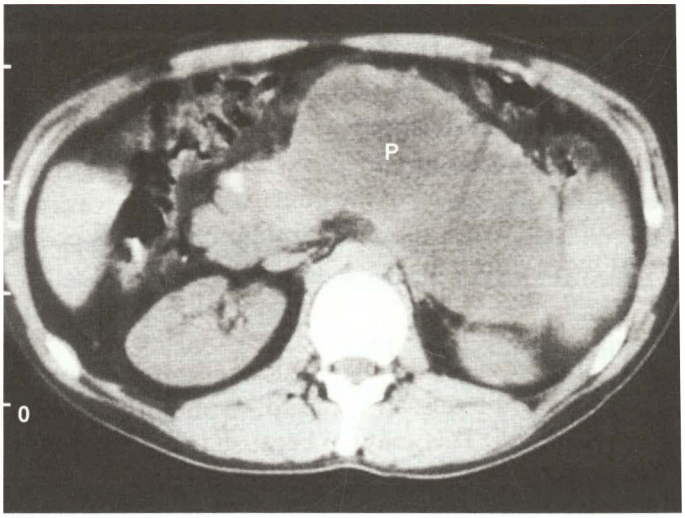


Fig. 25.132: CT scan showing infected necrosis

PEARLS OF WISDOM

Early aggressive fluid replacement is the key in the management of acute pancreatitis.

- Even though management principles of any acute abdomen includes 'nil per oral' so as to give rest to the part, in cases of pancreatitis it has been divided broadly into 3 categories. It is desirable to know the treatment followed for these categories. It has been given in Table 25.10 and Key Box 25.34.
- Those of you who find it difficult can remember at least few fundamentals of managing acute pancreatitis. They are given in the form of simple headlines: **ABCDEF**

A: Aspiration with Ryle's tube, to give rest to the pancreas (may be for one or two days).

B: Blood transfusion if Hb% is low, or albumin and amino acids if proteins are low.

C: Charts—increasing pulse, increasing temperature indicates pancreatic abscess which needs laparotomy and drainage.

Table 25.9 Prognostic indicators/severity predictors

Ranson score	Glasgow Scale
ON ADMISSION	
Age > 55 years	Age > 55 years
WBC count > 16 × 10^9/L	WBC count > 15 × 10^9/L
Blood glucose > 10 mmol/L	Blood glucose > 10 mmol/L
LDH > 700 units/L	Serum urea > 16 mmol/L
AST > 250 Sigma Frankel unit/unit	PaO_2 < 60 mm Hg
WITHIN 48 HOURS	
Increase in blood urea nitrogen levels > 5 mg%	Serum calcium < 2 mmol/L
Drop in haematocrit > 10%	Serum albumin < 32 g/L
Arterial oxygen saturation (PaO_2 < 60 mmHg)	LDH > 600 units/L
Serum calcium < 2 mmol/L	AST/ALT > 600 units/L
Base deficit > 4 mmol/L	
Fluid sequestration > 6 L	

Table 25.10 Management principles of acute pancreatitis

	Acute oedematous (Mild—80%)	Necrotising pancreatitis (Sterile necrosis—10%)	Infected necrosis (Very severe—5%)
Admission	Acute ward with monitoring of vitals	Intensive care unit	Intensive care unit
IV fluids/ hypotension	**Early correction of hypotension,** hypovolaemia—crystalloids	IV fluids and inotropic* support may be required	May require inotropes and vasopressors for a long period
Blood transfusion	Rarely required	May be required	Definitely required
Antibiotics	No antibiotics	Early antibiotic prophylaxis is required	Broad spectrum antibiotics—imipenem, ciprofloxacin, metronidazole, etc.
Oral/nutrition	Oral fluids, soft diet by 3–4 days, once pain and ileus settle down	If pain is still present even after 4 days, nasojejunal feeding to be done	Enteral feeding/nasojejunal feeding. If calories are not sufficient, total parenteral nutrition is required
Hypomagnesaemia/ hypocalcaemia	Usually will not be a problem	Correction is required	Correction is required
Oxygen	By nasal cannula/face mask may be required	Early ARDS—ventilatory support	Ventilatory support may be required
Role of surgery/ natural course	Majority of patients will not require surgery. If gall stones are impacted at sphincter of Oddi, endoscopic basketing and laparoscopic cholecystectomy should be done	By 10–15 days, may resolve completely or may develop into pseudocyst or infected necrosis which require surgery	Ideal time to operate is after 4 weeks when necrosis is demarcated well

* Dopamine 2–5 µg/kg/min can help in renal perfusion. 5–10 µg/kg/min acts as an inotrope and 10–20 µg/kg/min as a vasopressor. Noradrenaline 50–200 ng/kg/min as a vasopressor.

D: Drugs: Prophylactic antibiotics—used in cases of severe pancreatitis for prevention of local or general complication. IV cefuroxime or imipenem or ciprofloxacin with metronidazole are given. **Low molecular weight dextran** (lomodex) 500 ml can be used to increase renal perfusion. Alternately, dopamine 2 µg/kg/min can be given IV which helps in renal perfusion (in case of oliguria).

KEY BOX 25.34

INDICATIONS FOR SURGERY IN ACUTE PANCREATITIS

- Infected necrosis
- Pancreatic abscess
- Diagnosis is in doubt—perforated viscus cannot be ruled out.
- Complications such as massive bleeding not responding to conservative treatment.
- Cholangitis not responding to treatment.

E: Exploratory laparotomy, **only when diagnosis is in doubt,** when patient is **not improving** or when there is a complication of pancreatitis such as pancreatic abscess, fistula or necrosis. With the advent of wonderful imaging techniques, surgical intervention is rarely being done in the initial phase of acute pancreatitis. In early cases, pancreas should not be handled. Peritoneal lavage is done followed by insertion of tube drain. Lavage has shown some benefit. In cases of infected necrosis, necrosectomy is done.

- The wound can be left open as laparotomy or with mesh or with zip.

F: Fluid should be given early. Rapid infusion of 3–4 litres of Ringer lactate is used to treat hypovolaemic shock. Plasma or albumin may also be given.

- Factors predicting severe pancreatitis (Key Box 25.35).

KEY BOX 25.35

IDENTIFICATION OF SEVERE PANCREATITIS

- Age : > 70 years
- Obesity : BMI > 30 kg/m^2
- Pleural effusion : Present
- CT necrosis : > 50
- CT severity index : CTSI
- Persistent : Organ failure

PEARLS OF WISDOM

In cases of infected necrosis proved by CT-guided FNAC and Gram stain, necrosectomy should be done.

PANCREATIC NECROSECTOMY

(Figs 25.133 to 25.139)

- Ideally done 4–6 weeks later when sepsis is still present.
- CT-guided FNAC/bacterial culture is a must.
- A midline laparotomy is done
- Lesser sac is entered
- Thorough debridement of the dead tissue is done.
- All fluid and tissue should be sent for aerobic and anaerobic culture.
- Blunt dissection rather than sharp dissection is done to minimise bleeding.
- Antibiotics of choice should be carbapenems and quinolones.
- Ileostomy should be done for retrocolic necrosis
- Cholecystectomy should be done for gall stone pancreatiti cases.
- Perfect haemostasis should be achieved using manua compression, sutures and ligatures.
- In cases of early necrosectomy, lesser sac is packed reoperation done after 48 hours—zipper closure of the abdomen is used.
- Continuous lavage of the lesser sac and retroperitoneum is done.
- Other alternate methods are closed packing.
- Today all these cases are done through minimally invasive methods.
- Endoscopic necrosectomy, retroperitoneoscopic necrosectomy is also done.

Fig. 25.133: Acute necrotising pancreatitis. Patient had high temperature. Nasojejunal tube feeding is being done

Fig. 25.134: Acute necrotising pancreatitis with massive upper GI bleeding. Patient had high temperature, angiogram revealed pseudoaneurysm. He underwent therapeutic embolisation followed by necrosectomy

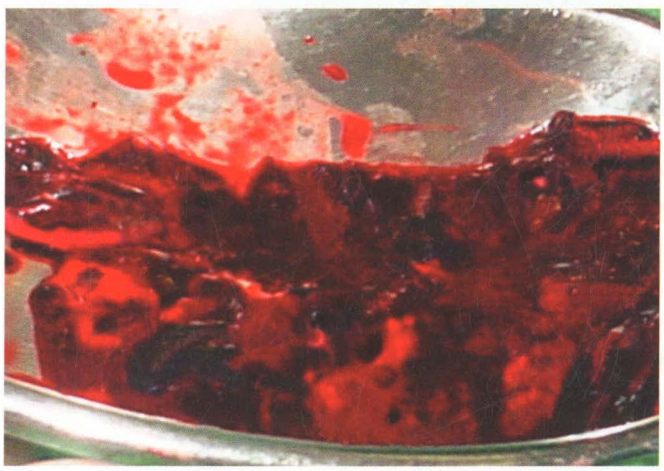


Fig. 25.135: Acute necrotising pancreatitis with massive upper GI bleeding. Patient had haematemesis of one litre of blood with clots

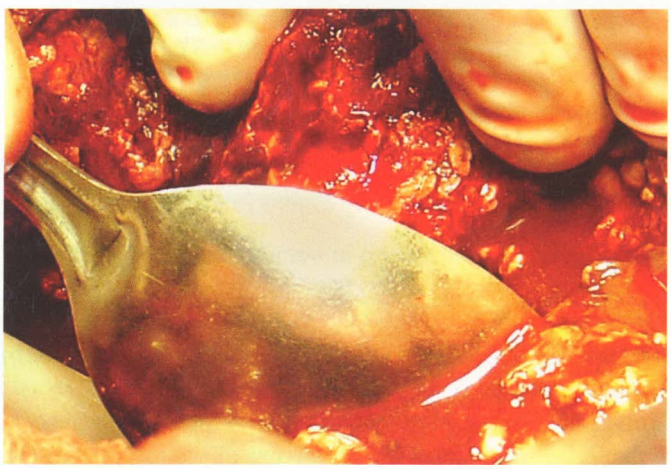


Fig. 25.136: You can see necrosectomy being done. It was done through lesser sac. Significant blood loss was present. Fingers are used to separate necrosis and it is removed with a spoon

Fig. 25.137: After necrosectomy, pancreatic bed is washed with saline and tubes or sump drains are put so as to be able to irrigate in the postoperative period

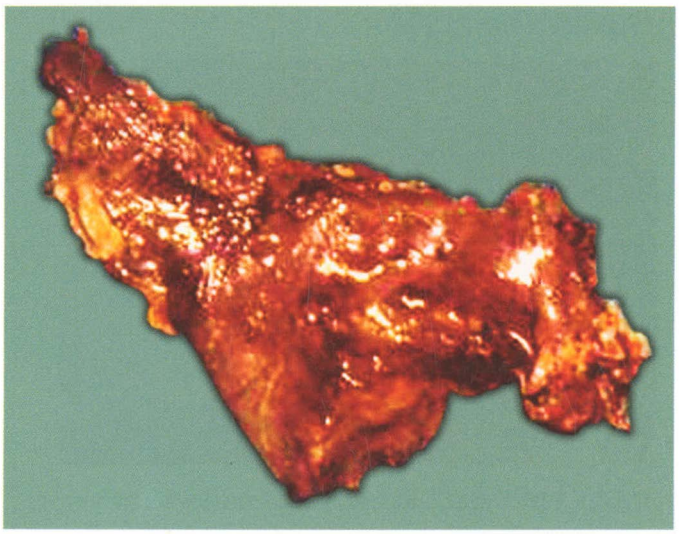


Fig. 25.138: Necrosectomy specimen

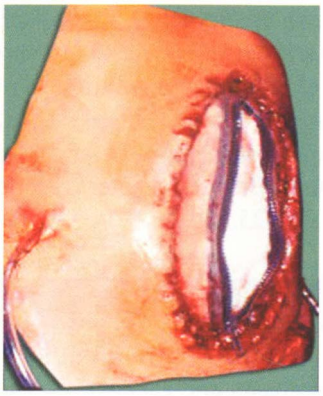


Fig. 25.139: After necrosectomy, it is better to perform zip laparotomy so that it can be opened any number of times for peritoneal toilet (*Courtesy:* Dr BH Ananda Rao, Prof and Head, Department of Surgery, Dr Siddhartha Bhandary and Dr Prashanth Shetty, Associate Professors, KMC, Manipal)

Treatment of gall stone pancreatitis (Key Box 25.36)

If gall stone is obstructing the ampulla of Vater and causing pancreatitis, endoscopic sphincterotomy may be done within days. It has shown some benefit to the patients. After 2–3 weeks, once the jaundice subsides, cholecystectomy is recommended.

KEY BOX 25.36

ACUTE PANCREATITIS

- Lipase is a more reliable test because it is more pancreas-specific and remains elevated for long after onset of symptom.
- However, amylase is widely used
- It is not necessary to demonstrate CBD stone to prove the diagnosis of gall stone pancreatitis. Often stone has passed into duodenum.

COMPLICATIONS OF ACUTE PANCREATITIS (Table 25.11)

Systemic complications

1. **Shock**

- Hypovolaemia and hypoperfusion are the major factors responsible for renal failure. Due to collection of large

Table 25.11 Complications of acute pancreatitis

Local	General
• Pancreatic necrosis (sterile)	• Pulmonary—ARDS
• Pancreatic necrosis (infected)	• Cardiac—shock, arrhythmias
• Pancreatic abscess	• Renal—renal failure
• Pancreatic ascites	• Gastro—ileus, intestinal colonic necrosis
• Pancreatic pseudocyst	• Metabolic—hypercalcaemia, hypoglycaemia, hyperlipidaemia
• Peritoneal fluid collection	• Haematological
• Pleural effusion	
• Pseudoaneurysm	
• Partial/splenic vein thrombosis	

amount of fluid in the **third space**—peritoneal cavity, pleural cavity and extravascular space, shock occurs. Fluid replacement with blood or albumin should be done at appropriate time to treat the shock.

- Electrolyte abnormalities should be corrected.

2. **Respiratory insufficiency:** Factors responsible for this are given in Key Box 25.37.

KEY BOX 25.37

RESPIRATORY FAILURE

- Abdominal distension and elevation of diaphragm.
- Intravascular coagulation in the lung.
- Lecithin present in the pulmonary surfactant is altered due to lecithinase resulting in defective capillary alveolar exchange.
- Defective ventilation caused due to pain.
- Left-sided pleural effusion not responding to treatment.

Measurement of arterial blood gas values and administration of oxygen is enough in the initial stages. In late stages, pulmonary insufficiency needs to be treated with ventilatory support.

3. **Hypocalcaemia** needs to be treated with calcium IV. It is due to hypoalbuminaemia and due to calcium soap.
4. **Pleural effusion** is treated by pleural tap (ultrasound-guided), if it is symptomatic.
5. **ARDS, MODS:** Some mediators such as phosphatase damage alveolar membrane of lungs causing 'ARDS'. It manifests as respiratory failure.

Local complications

1. **Pancreatic abscess** (Fig. 25.140)

- It develops after 3–4 weeks of pancreatitis. Secondary infection in a pseudocyst results in pancreatic abscess. It usually points out on the left flank.
- It has to be drained by CT-guided aspiration.
- Laparoscopy may be necessary not only for the diagnosi but also as a therapeutic means for removal of necroti pancreas.
- Otherwise, open drainage of the abscess is required.

2. **Pseudocyst of pancreas** ***(vide infra)***

- This complication is encountered after 2nd week following an attack of acute pancreatitis.
- It is seen in about 20% of the patients.

3. **Perforation** of colon or stomach
4. **Pseudoaneurysm** resulting in massive upper gastro-intestinal or lower gastrointestinal bleeding. Bleeding into the pancreatic duct is called **haemosuccus pancreaticus**.

- This condition occurs due to enzymatic digestion of the blood vessels in the vicinity of pancreas. Thus splenic artery, gastroduodenal artery, etc. are commonly involved.
- It has very high mortality. Prompt angiography followed by embolisation is the treatment.
- Otherwise, laparotomy, ligation of pseudoaneurysm with or without intracystic ligation of the bleeders is the treatment (Figs 25.141 to 25.143).

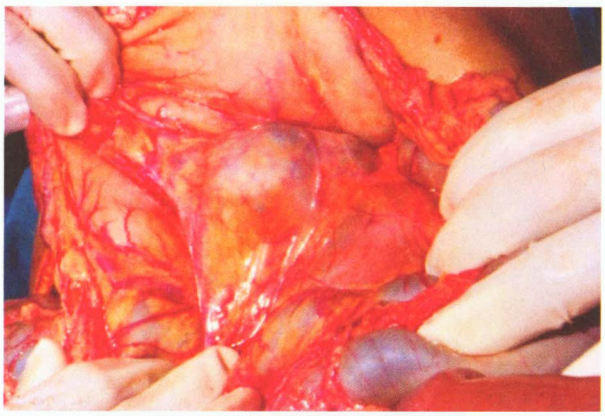


Fig. 25.141: Gastroduodenal pseudoaneurysm at surgery

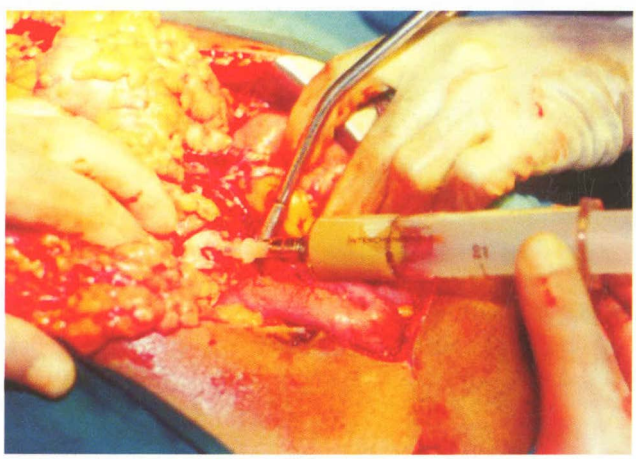


Fig. 25.140: Aspiration of pus from pancreatic abscess

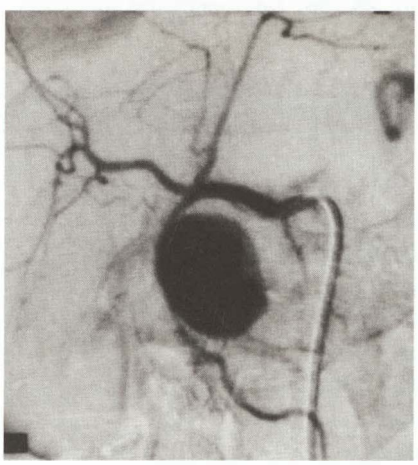


Fig. 25.142: Diagnosis by angiography

Fig. 25.143: As a cause of massive upper gastrointestinal bleeding (*Courtesy:* Dr Annappa Kudwa, Professor of Surgery, KMC, Manipal)

PSEUDOCYST OF PANCREAS

Definition

Collection of amylase-rich fluid in the lesser sac, due to pancreatic pathology. Fluid collection in the first 4 weeks is an acute fluid collection. After 4 weeks, it becomes an acute pseudocyst.

PEARLS OF WISDOM

It is called pseudocyst because it has no epithelial lining.

- Fluid is enclosed by a wall of fibrous granulation tissue and is called a pseudocyst.

Aetiology

1. Following an attack of **acute pancreatitis**, it usually appears after 4 weeks, as upper abdominal swelling.
2. **Blunt injury** of abdomen causing a ductal disruption wherein the pancreatic duct in the region of body is crushed against vertebral body results in a pseudocyst.
3. Some cause of **chronic pancreatitis** may be associated with pseudocyst.

Locations of pseudocyst

1. Between stomach and transverse colon
2. Between stomach and liver
3. Behind or below the transverse colon

D'EGIDIO CLASSIFICATION OF PSEUDOCYST

Type I. Acute postnecrotic pseudocyst that occurs after **an episode of acute pancreatitis** and is associated with **normal duct anatomy** and rarely communicate with pancreatic duct.

Type II. Post necrotic pseudocyst that occur after an **episodic of acute on chronic** pancreatitis and have a diseased but **not strictured** pancreatic duct and **there is after—communication between the duct and pseudocyst.**

Type III (also called retention cyst) **occur in chronic pancreatitis,** uniformly associated with a duct stricture and a communication between duct and pseudocyst.

Clinical features

1. Tensely cystic mass in the epigastrium, umbilical region or in left hypochondrium. Tensely cystic mass feels firm on palpation. Classically upper border of the mass is not felt.
2. Does not move with respiration because it is retroperitoneal in location.
3. It may have transverse mobility
4. It does not fall forwards
5. Percussion: It gives a **resonant note** because of stomach or intestine anterior to it.
6. **Transmitted pulsations** from aorta can be felt
7. If a Ryle's tube is passed, it can be felt over the swelling. It is called **Baid sign**.
8. Depending upon the tension within the cyst, it can be tender or nontender.

Investigations

1. USG/CT: It can detect size, location of cyst and wall thickness. Confirm that there is no neoplasm of the pancreas before doing cystogastrostomy (Fig. 25.144).
2. Barium meal stomach: In a lateral picture, the stomach is pushed anteriorly and increased vertebrogastric interval is seen (not done nowadays, Fig. 25.145).
3. ERCP may demonstrate communication of the cyst with the duct (routinely not done).

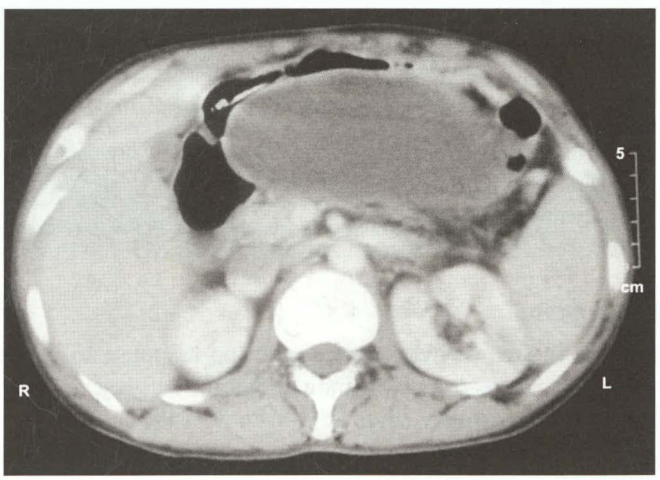


Fig. 25.144: CT scan: Classical pseudocyst pushing the stomach anteriorly—cystogastrostomy is ideal treatment

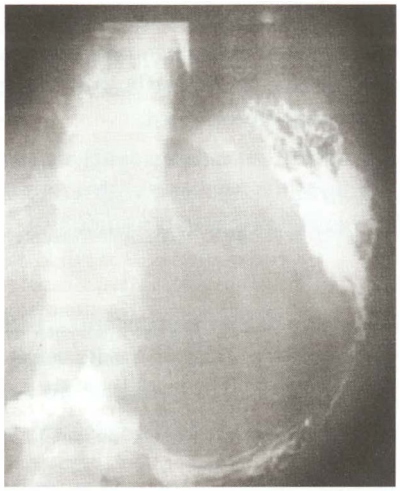


Fig. 25.145: Large pseudocyst behind the stomach demonstrated by barium meal

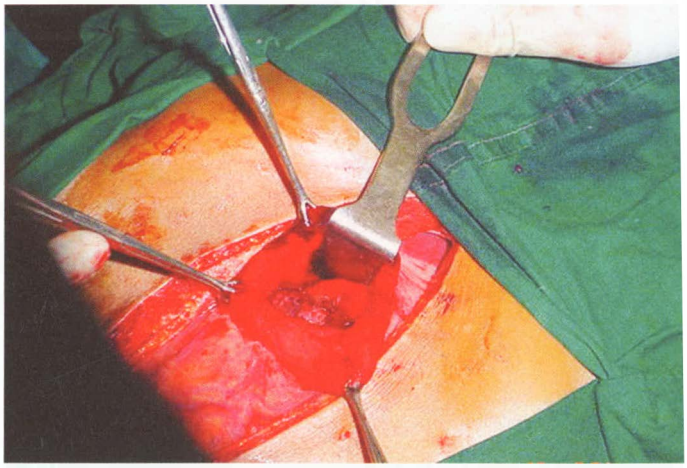


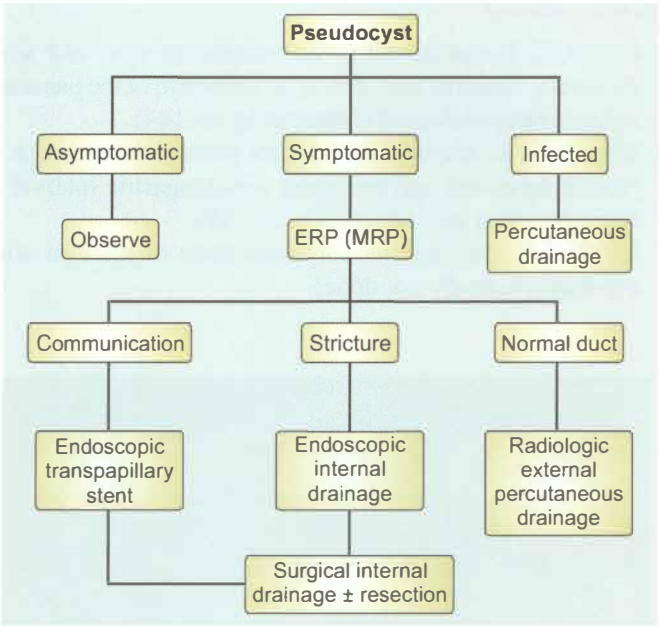
Fig. 25.146: Posterior gastric wall is sutured to anterior cyst wal after creating a stoma of 6 cm

Treatment

I. Conservative line of treatment

- Majority of the pseudocysts following acute pancreatitis resolve spontaneously within 3–4 weeks.
- Hence, regular ultrasound examination is done to observe the pseudocyst.

Flowchart of management of pseudocyst



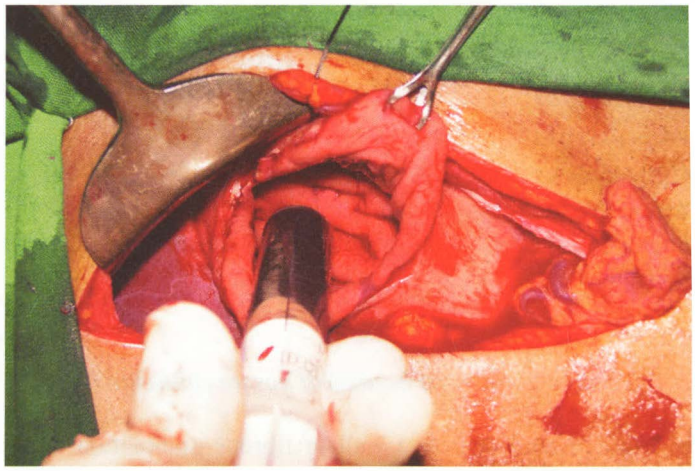


Fig. 25.147: Cystogastrostomy

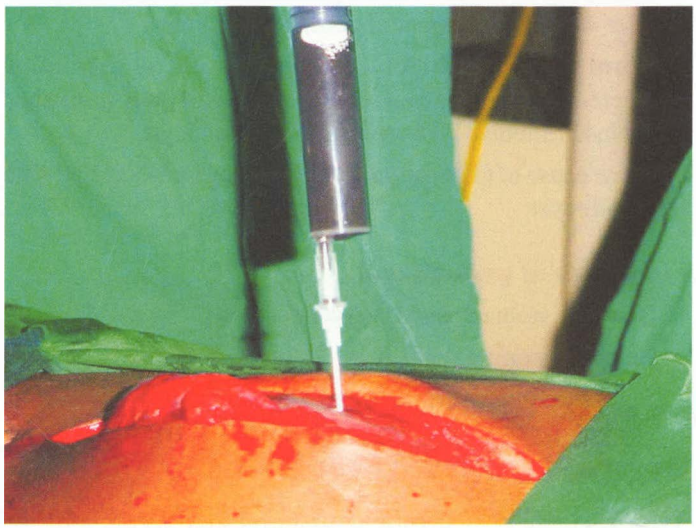


Fig. 25.148: Aspirated fluid is dark in colour, rich in enzymes—'engine oil' appearance

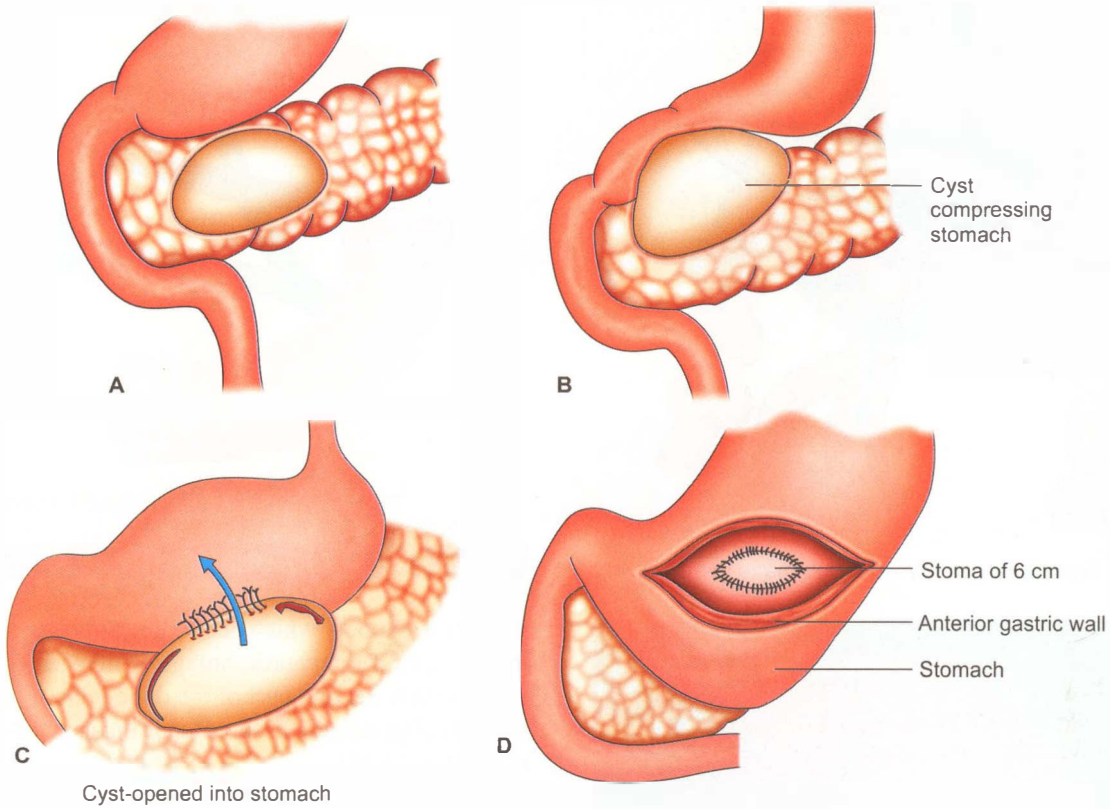
II. Surgery

Increase in size of cyst, severe pain, no response to conservative line of treatment are indications for surgery.

1. **Cystogastrostomy** (Figs 25.146 to 25.149)

Indications: Pseudocyst in relation to head and body of pancreas.

- **Timing:** Surgery is done **after 6 weeks** because that is the time required for the wall to become fibrous.



Figs 25.149A to D: Steps of cystogastrostomy (*Courtesy:* Miss Vidushi, MBBS student, KMC, Manipal)

- **Size** of the cyst should be at least **6 cm**.
- **Procedure:** Anterior gastrostomy is done and an incision in the posterior wall of stomach opens into the cyst cavity. The contents are drained, opening is enlarged and cut end of stomach in posterior wall is sutured to cut edge of cyst wall. After one week, the cyst collapses. For reasons not known, the food does not enter the cyst cavity. **Size of cystogastrostomy stoma is about 6 cm. This procedure can also be done by endoscopic method.**

2. **Distal pancreatectomy:** Cyst confined to the tail of the pancreas is treated with removal of the tail and the cyst.
3. **Cystojejunostomy:** By using Roux-en-Y loop can be done, **for large cysts** by suturing jejunal loop to the cyst in the most dependent area (Figs 25.150 to 25.152).

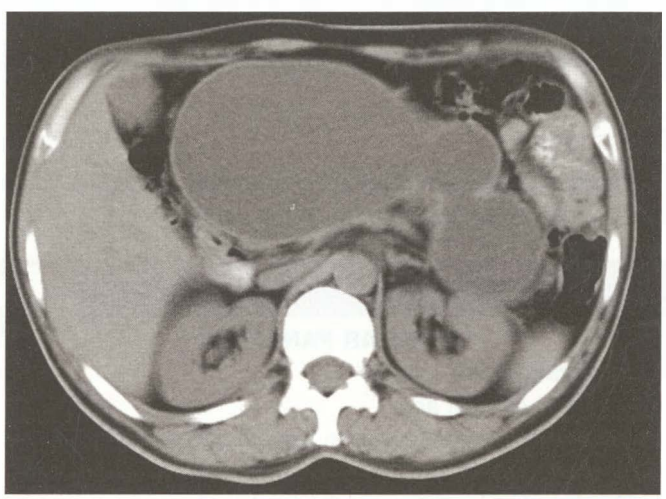


Fig. 25.150: CT scan showing large pseudocyst with multiple pockets. Cystojejunostomy is the ideal treatment

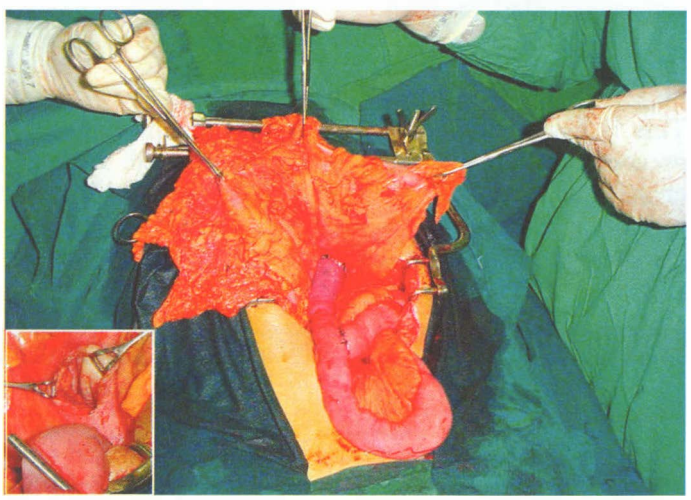


Fig. 25.151: Roux-en-Y cystojejunostomy at surgery, inset shows the opened cyst wall which is thick

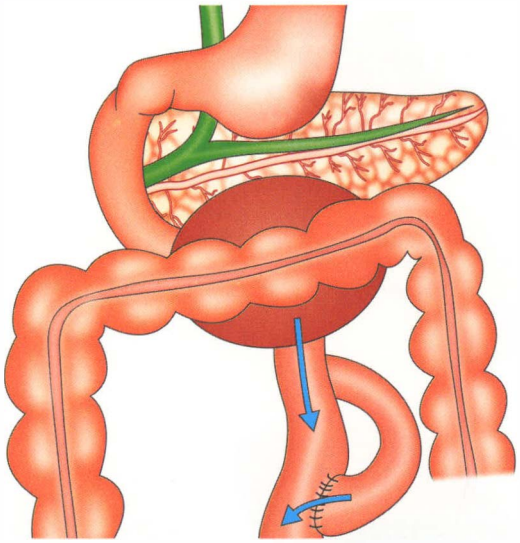


Fig. 25.152: Cystojejunostomy—diagrammatic representation

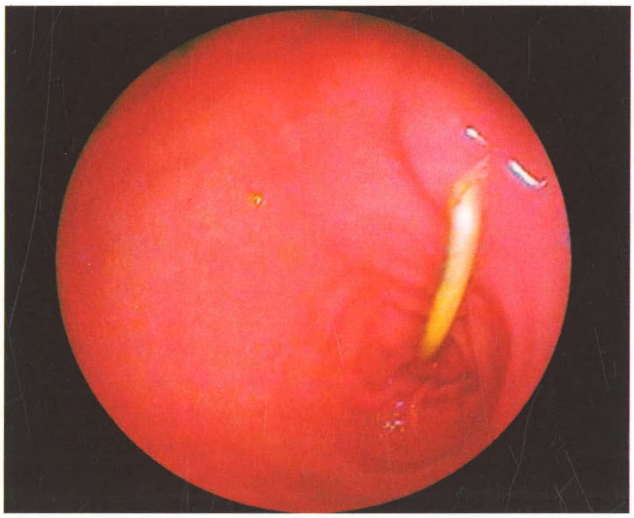


Fig. 25.153: Endoscopic cystogastrostomy—ideal to drain acute pseudocysts. Inset—after draining a nasobiliary catheter can be left in the cyst cavity (*Courtesy:* Dr Filipe Alvares, Medical Gastroenterologist, KMC, Manipal)

III. Endoscopic drainage

This is easy, less invasive and ideal in draining acute pseudocysts. This procedure can also be repeated. However, chances of introducing infection are present. Endoscopic method of draining the pseudocysts is also being done for chronic cysts (Fig. 25.153).

CONGENITAL ANOMALIES OF THE PANCREAS

Embryological anatomy

- Pancreas is formed by fusion of ventral and dorsal buds. This will occur by 6 weeks. Malrotation of ventral bud causes annular pancreas.
- Ventral bud forms head and uncinate process (Fig. 25.154).

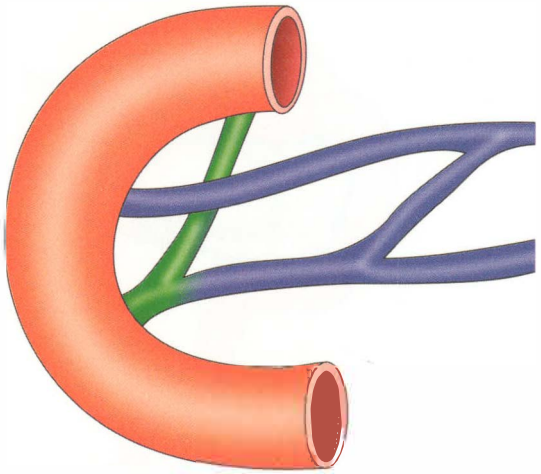


Fig. 25.154: Pancreatic ductal anatomy

ANNULAR PANCREAS

It is a rare anomaly which occurs due to persistence of a portion of the ventral pancreatic anlage which fails to rotate. As a result of this, second part of the duodenum is surrounded by a thin rim of pancreatic tissue. Hence the name, annular pancreas (Key Box 25.38).

Associated anomalies

Intrinsic duodenal atresia or stenosis.

Clinical features

- **Neonatal type:** It manifests early in life. It produces symptoms of acute intestinal obstruction with vomiting and inability to take food.
- **Adult type:** It manifests after the age of 20. Vomiting is bile-stained. Due to stasis in the pyloric antrum, features of duodenal ulcer may be present.

Investigations

- **Plain X-ray abdomen:** Double bubble appearance occurs due to dilated stomach and dilated proximal duodenum.
- **Barium meal:** X-ray can demonstrate obstruction to second part of duodenum.

Treatment

- **Duodenoduodenostomy** is the treatment of choice. Otherwise, duodenojejunostomy can be done.

KEY BOX 25.38

ANNULAR PANCREAS

- Persistent ventral 'anlage'
- Neonatal or adult age
- Barium meal—obstruction to second part of duodenum
- Plain X-ray—'double bubble'
- Duodenoduodenostomy is the treatment
- Never divide the annulus

PEARLS OF WISDOM

It is tempting to divide the thin pancreatic rim. This should not be done as it will result in a pancreatic fistula.

Differential diagnosis

1. Pyloric stenosis
2. Wilkie's disease (chronic duodenal ileus)

ECTOPIC PANCREAS

- This condition is not uncommon
- Occasionally, at laparotomy for unrelated conditions, a soft to firm irregular tissue or nodule, is found on the intestinal surface.
- Biopsy of this may come later as ectopic pancreas. Most of them are asymptomatic.
- It may be found in the submucosa of the stomach, duodenum, small intestines, Meckel's diverticulum or in the hilum of the spleen.

Complications

1. In the stomach: It may undergo cystic degeneration.
2. In the intestine: It can cause intussusception. Sometimes, it may be the source of gastrointestinal bleeding.
3. In the Meckel's diverticulum: Any of the complications mentioned above.

CONGENITAL CYSTIC FIBROSIS

- It is inherited as an autosomal recessive disorder.
- It is a generalised dysfunction of exocrine glands resulting in defective mucus secretion.
- Malabsorption due to pancreatic insufficiency is a feature.
- Pulmonary disease due to bronchiolitis occurs later (Key Box 25.39).

KEY BOX 25.39

VISCID MUCUS

- Meconium ileus
- Cystic fibrosis of pancreas
- Respiratory tract infection
- Increased Na^+ loss in the sweat

Pathology

The viscid mucin which is produced results in obstruction of the ducts and ductules. Stasis of pancreatic secretions, alveolar rupture due to increased pressure take place later. As a result of alveolar rupture, pancreatic enzymes leak outside resulting in pancreatitis.

Clinical features

1. **At birth:** Meconium ileus or meconium peritonitis is an important manifestation of cystic fibrosis of pancreas.
2. **In infants**
   - Recurrent respiratory tract infection in the form of bronchiolitis and bronchiectasis results in cough with expectoration and dyspnoea.
   - Emaciation, steatorrhoea. Stools are pale and sticky.
3. **Older children**

   Steatorrhoea, gross emaciation and wasting are the features. Due to poor nutritional status, cirrhosis of the liver with portal hypertension can be a feature.
4. **In adult**

   Lucky survivors will suffer from gross wasting, diabetes, bronchiectasis, cirrhosis of the liver, sialoadenitis and choroiditis.

Investigations

- Normal contents of the electrolytes in the sweat: Na^+—70 mmol/L, Cl^-—60 mmol/L, K^+— 20 mmol/L.
- In these patients, sodium excretion may be three to four times more than normal.

Treatment

Only symptomatic

I. Nutritional support
- Fat intake should be low, protein should be increased.
- Pancreatic enzyme preparations (5 to 10 g) are given two to three times per day, to supplement pancreatic enzymes.

II. Control infection

Respiratory tract infection is treated with antibiotics, bronchodilators, mucolytic agents, etc.

III. Role of surgery

Indicated in meconium ileus to relieve intestinal obstruction.

PANCREATIC DIVISUM AND DUCTAL ANOMALIES

- Pancreatic divisum is the most common congenital abnormality of the pancreas. It is found in 5–10% of patients.
- It is caused by failure of fusion of the dorsal and ventral portions of the developing pancreas. In the majority of patients, this anomaly is of no clinical importance.
- In a certain subset of patients, however, estimated to be approximately 5–20%, pancreatic divisum is a clinically important cause of abdominal pain, acute recurrent pancreatitis or chronic pancreatitis.
- **The frequency of pancreatic divisum in patients with pancreatitis in recent studies is approximately 10%** (Key Box 25.40).

KEY BOX 25.40

PANCREATIC DIVISUM

- It is one of the uncommon causes of acute pancreatitis.
- In 10% of patients. Majority of pancreas drains through duct of Santorini through lesser papilla.
- Outflow obstruction—treated by sphincteroplasty of minor papilla.

Pathophysiological consequences

The cause of pancreatitis in this common anomaly is hypothesised to be caused by minor papillary insufficiency caused by papillary fibrosis and subsequent stenosis.

Diagnosis

- The diagnosis is established by ERCP.
- MRCP is a noninvasive and accurate method of establishing the diagnosis. However, a negative MRCP does not exclude the diagnosis of divisum as the concordance with ERCP ranges from 50% to more than 70%.
- The accuracy of the MRCP may be improved by administration of secretin.

Treatment

- In patients with minimal symptoms, treatment with pancreatic enzymes may reduce pancreatic secretion and hence, pain. Pain management referral may be necessary.
- Patients with acute pancreatitis are estimated to benefit from either surgical minor sphincteroplasty or endoscopic minor papillotomy.

A few anomalies of pancreatic duct are given below (Figs 25.155A to E)

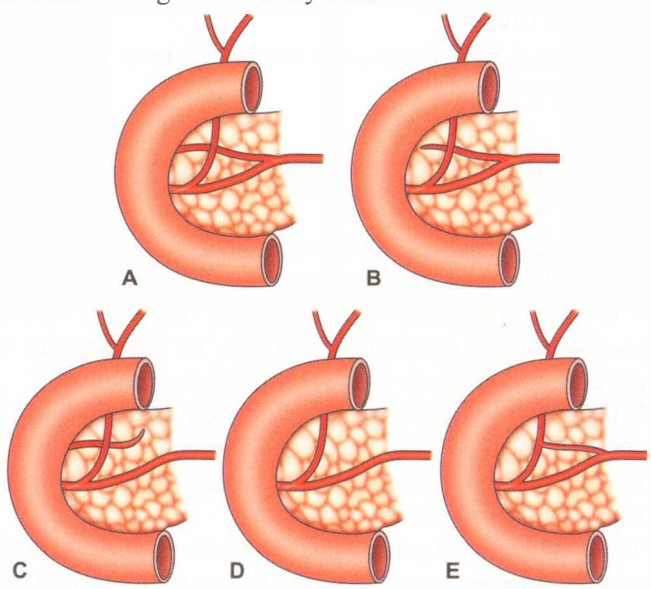
A. Normal anatomy

B. Terminal end of accessory duct fibrosed

C. Blind ending of accessory duct

D. Absent accessory duct

E. Blind end of accessory duct with main duct communication



Figs 25.155A to E: Anomalies of pancreatic duct

MISCELLANEOUS

PANCREATIC FISTULA

Most common after **pancreatic trauma** (Fig. 25.156).

Causes

1. **External fistulae** occur due to operative injury to the pancreas or due to a pancreatic anastomotic leak.
   - Injury to the tail of pancreas during splenectomy or adrenalectomy.
   - Injury to the head and body during radical gastrectomy.
   - Pancreaticojejunostomy for chronic pancreatitis or following Whipple's operation can also give rise to fistula in the postoperative period.
   - External drainage of an infected pseudocyst.
2. **Internal fistulae:** It can occur following a blunt injury abdomen wherein the neck of the pancreas is crushed against lumbar spine resulting in injury. Internal fistulae can communicate with pleural space resulting in a pancreaticopleural fistula.

Clinical presentation

- In many cases, the patients present with a discharge of straw-coloured fluid from the drain site in the postoperative period.
- Internal fistula can manifest in a totally unexpected manner, sometimes, as a case of pleural effusion.

Investigations

- **Amylase** levels in the pleural fluid, peritoneal fluid and in the discharge will be high.

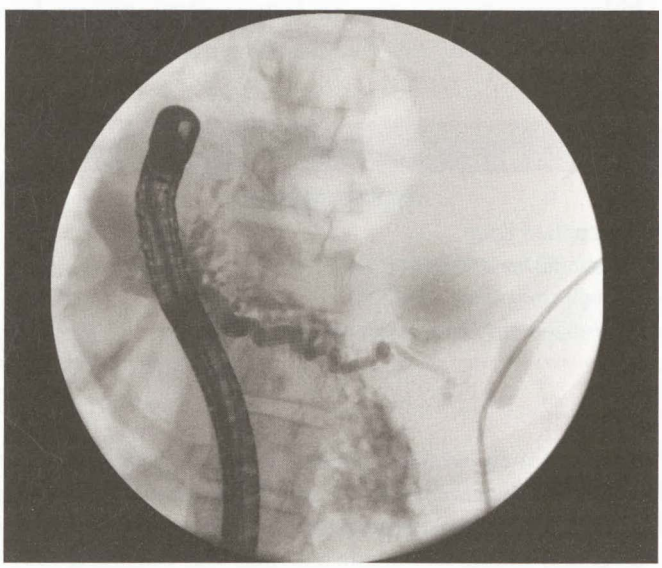


Fig. 25.156: Pancreatic fistula following acute pancreatitis

- Abdominal ultrasound is done to rule out a pseudocyst of the pancreas.
- **ERCP:** It can demonstrate leakage of the dye from the pancreatic duct into the surrounding area or along the fistulous tract and proximal obstruction, if any.
- **CT fistulogram** can define the exact site of communication to pancreas. It can define the length of the fistula also.
- **CT angiogram** is indicated in cases of bleeding associated with pancreatic fistula.

Treatment

1. Conservative treatment

- In majority of the cases following surgery, the fistulous discharge stops within one to three weeks of time. During this period, the skin is protected by application of zinc oxide cream. Electrolytes have to be checked frequently.
- Injection octreotide in the dose of 50–100 mg, 8th hourly will help in decreasing fistula output by more than 80–90%.

2. Surgical treatment

- If the **fistula persists** in spite of conservative treatment, **fistulectomy** with removal of involved part of the pancreas and body or tail has to be done.
- In very difficult cases (plastered abdomen) or in fistulas on the anterior abdominal side, **fistulogastrostomy** can also be done.
- Pancreaticojejunostomy is another option.

Complications of pancreatic fistula

1. Secondary **infection**
2. **Massive bleeding:** It occurs due to digestion of elastin fibres of blood vessels resulting in pseudoaneurysms. Morbidity is high.
3. **Ramification** which means branching pattern of the fistula renders the surgery difficult.
4. **Bronchopancreatic fistula**—difficult to treat

CLINICAL NOTES

A patient was admitted to the hospital with acute breathlessness. He was found to have left-sided pleural effusion (massive). He had blunt injury abdomen 2 months back during which time laparotomy and closure of a proximal jejunal perforation had been done. There was no obvious sign of pancreatic injury. Aspiration of pleural fluid revealed high amylase. It was a case of pancreaticopleural fistula.

WHITE BILE

- It is a misnomer (Fig. 25.157)
- In long-standing cases of obstruction to the CBD, the bile in the CBD gets absorbed and is replaced by mucus secreted from the CBD.

Fig. 25.157: White bile suggestive of severely impaired liver cell function

- It is not white but straw coloured
- It is not bile but is mucus

Significance

- It indicates a long-standing obstruction.
- It has to be relieved as an urgent procedure.
- White bile is seen in:
  - Long-standing stricture of the CBD
  - Due to the stones in CBD
  - Rarely, seen in periampullary carcinoma.

PANCREATIC ASCITES

Definition

- Accumulation of 'enzyme rich' pancreatic exudate in the peritoneal cavity is called pancreatic ascites.
- It is a protein-rich, noninfected fluid with protein levels greater than 25 g/L.

Causes

- Acute pancreatitis mainly alcoholic
- Chronic pancreatitis
- Trauma to pancreas
- Ruptured pseudocyst

Pathogenesis

- Disruption of ductal system of pancreas followed by spread of enzyme-rich fluid both anteriorly and posteriorly results in pancreatic ascites.
- Anterior rupture results in ascites.
- Posterior spread results in pleural effusion.

Clinical features

- History suggestive of pancreatitis, gross abdominal distention.
- Shifting dullness
- Breathlessness—due to pleural effusion.

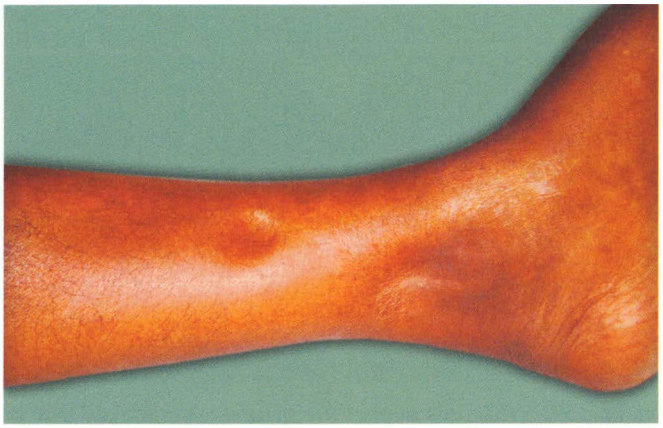


Fig. 25.158: Pedal oedema—severe hypoalbuminaemia

Fig. 25.159: Clinical presentation—tense ascites

Investigations

- Serum amylase, may be elevated due to reabsorption across parietal membrane.
- Ultrasound/CT: It can detect fluid, pancreatic duct and guide aspiration.
- ERCP: May slow ductal communication.

Treatment

- Repeated **tapping**—follow with **albumin infusion.** Most of the cases are associated with hypoalbuminaemia (Figs 25.158 and 25.159).
- Octreotide to decrease pancreatic secretion.
- Total parenteral nutrition
- ERCP—pancreatic duct stenting
- Surgical: If there is no response by 2–3 weeks, resection (for tail lesion) or drainage for body lesion is indicated.

ISLET CELL TRANSPLANTATION

- This is emerging as a fine treatment for diabetes and chronic pancreatitis after pancreatectomy.
- Many patients have undergone islet cell transplantation with minimal complications (*see* Chapter 49).

WHAT IS NEW IN THIS CHAPTER? /RECENT ADVANCES

- All the topics have been updated.
- Treatment of acute pancreatitis has been given in more detail with more illustrations.
- Islet cell transplantation, percutaneous cholecystostomy and pancreatic divisum have been added.

MULTIPLE CHOICE QUESTIONS

1. Which of the following statements is false for pseudocyst of pancreas?
A. It occurs in lesser sac
B. Cystogastrostomy is usually the treatment of choice
C. Better to operate after 2 weeks
D. It has no epithelial lining

2. Which of the following is not the cause for gall stone formation?
A. Saint's triad
B. Haemolytic anaemia
C. Infection
D. Gastrojejunostomy

3. Gall bladder carcinoma has following features *except:*
A. It is squamous cell carcinoma
B. It does cause jaundice often
C. It does not respond to radiation
D. Prognosis is very poor

4. Most ideal and quick method for the diagnosis of gall stones is by:
A. Endosonogram B. MRI scan
C. CT scan D. Ultrasound

5. Following is not the feature of acute cholecystitis:
A. It causes pain in the right hypochondrium
B. If it perforates, gas under diaphragm is detected by percussion
C. Shoulder pain can occur
D. Acalculous cholecystitis is due to hypotension

6. Following is the most important sign of acute cholecystitis:
A. Pain in the right shoulder
B. Intercostal oedema and tenderness
C. Positive Murphy's sign
D. Hyperaesthesia of the abdominal wall

7. Cystic duct joins supraduodenal CBD in what percentage of the cases?
A. 80% B. 90%
C. 70% D. 95%

8. Porcelain gall bladder has following features *except:*
A. It is calcification of the gall bladder
B. Plain X-ray is not very useful to detect this
C. CT scan will detect this
D. It may be associated with carcinoma gall bladder

9. Which one of the following is true for choledochal cyst?
A. Acquired dilatation
B. Not premalignant
C. Type I cyst is the most common variety
D. Does not cause pancreatitis

10. Which one of the feature is true in gall stone ileus?
A. Obstruction is in terminal ileum
B. Stone reaches ileum via ampulla of Vater
C. Obstruction and features of perforation of gall bladder are found
D. Cholecystectomy and extraction of the stones by enterotomy is done

11. Following are true for acalculous cholecystitis *except:*
A. It can be seen in septic shock due to hypotension
B. Some features of cholecystoses may also be found
C. *Salmonella typhi* can also give rise to acute acalculous cholecystitis
D. Rare to get chronic cholecystitis in Salmonella cholecystitis

12. Following are indications for common bile duct exploration *except:*
A. Palpable stones in the CBD
B. Dilated common bile duct
C. Jaundice
D. Thickened common bile duct

13. Following are risk factors for cholangiocarcinoma *except:*
A. Choledochal cyst
B. Caroli's disease
C. Primary sclerosing cholangitis
D. Biliary stricture

14. Which one of the following is true for carcinoma gall bladder?
A. It is common in males
B. Disease occur around 5th decade
C. Gall stone disease does not predispose to carcinoma gall bladder
D. Majority are adenocarcinoma

15. Caterpillar hump is an anomaly of what structure?
A. Cystic artery
B. Hepatic artery
C. Coeliac artery
D. Gastroduodenal artery

16. Portal veins is formed at:
A. Posterior to head pancreas
B. Posterior to body of pancreas
C. Anterior to neck of pancreas
D. Posterior to neck of pancreas

17. Following are true about anatomy of the pancreas *except:*
A. 30% is by the head of pancreas
B. 75% of islet cells are beta-cells producing insulin
C. 80–90% pancreatic tissue is exocrine pancreatic tissue
D. 5% of islet cells are A cells producing glucagon

18. To avoid spillage of cells after FNAC of the pancreatic head mass which is the best investigation?
A. Endoscopic biopsy
B. CT-guided biopsy
C. MRI-guided biopsy
D. Endosonographic biopsy

19. Which one of the following is true for pancreas divisum?
A. Dorsal pancreatic duct become accessory duct
B. It does not cause pancreatitis
C. MRCP is not an ideal investigation for diagnosis
D. Endoscopic sphincterotomy has a definitive role

20. Following are true for annular pancreas *except:*
A. It is more prevalent in children with Down syndrome
B. It surrounds 2nd part of duodenum
C. Duodenal obstruction causes vomiting in neonate
D. Gastrojejunostomy is the treatment of choice

21. The use of ultrasound in the diagnosis of acute pancreatitis is mainly to:
A. Rule out perforation
B. To look for oedematous pancreatitis
C. To rule out gall stones
D. To look for pancreatic stones

22. Role of CT scan in acute pancreatitis include following *except:*
A. To detect gall stones
B. Diagnostic uncertainty
C. To detect necrotizing pancreatitis
D. To find out the localized complication

23. Following are features of acute pancreatitis *except:*
A. Shock B. Cullen's sign
C. Grey Turner's sign D. Kehr sign

24. Which one of the following is not routinely recommended in severe pancreatitis?
A. Aggressive fluid resuscitation
B. Nasogastric feeding
C. ICU monitoring with oxygenation
D. Antibiotic prophylaxis

25. Which one of the following is true for pancreatic necrosectomy?
A. Usually done within 2–3 weeks of acute attack of pancreatitis
B. Gall stones—if present, should not be removed along with necrosectomy
C. Blunt dissection is the best technique
D. Feeding jejunostomy may not be useful

26. Sudden rise in platelet counts in acute pancreatitis suggests:
A. Pancreatic fistula
B. Bleeding
C. Portal vein thrombosis
D. Pancreatic necrosis

27. Following are true about pseudocyst fluid *except:*
A. Ultrasound can easily detect pseudocyst
B. CEA levels are usually above 400 ng/ml
C. High amylase levels
D. Inflammatory cells in the aspirate

28. Following are true for tropical pancreatitis *except:*
A. Starts in young age
B. High incidence of stone formation
C. High incidence of diabetes mellitus
D. Does not predispose to pancreatic cancer

29. The best surgical treatment for chronic pancreatitis with dilated duct is:
A. Frey's operation
B. Whipple's operation
C. Longitudinal pancreaticojejunostomy
D. Distal pancreatectomy

30. Following are the risk factors for carcinoma pancreas *except:*
A. Cigarette smoking
B. Hereditary pancreatitis
C. Chronic pancreatitis
D. Female sex

ANSWERS

1 C	2 D	3 A	4 D	5 B	6 D	7 A	8 B	9 C	10 A
11 D	12 D	13 D	14 D	15 B	16 D	17 D	18 D	19 D	20 D
21 C	22 A	23 D	24 B	25 C	26 C	27 B	28 D	29 C	30 D

26

Spleen

- Introduction
- Surgical anatomy
- Functions of the spleen
- Congenital abnormalities
- Rupture
- Complications of splenectomy
- ITP
- Hereditary spherocytosis
- Acquired autoimmune haemolytic anaemia
- Thalassaemia
- Sickle cell anaemia
- Splenectomy for other conditions
- Splenic artery aneurysm
- Hairy cell leukaemia
- OPSI
- Interesting 'most common'
- What is new?/Recent advances

Introduction

Diseases of the spleen and causes of enlargement of the spleen are a great concern for physicians. Dozens of differential diagnosis of splenomegaly are being taught in medicine. However, a surgeon's role in the treatment of an enlarged spleen is minimal. However, it is he **who does this 'magic' of removing** the spleen within minutes and saves the life of a patient with a ruptured spleen. A surgeon plays a major role by **treating hypersplenism with splenectomy**. In this chapter, only topics related to surgeon's interest are dealt with. Other topics are mentioned briefly.

SURGICAL ANATOMY (Fig. 26.1)

The spleen is an anatomically small organ, hidden underneath the **9**th to **11**th ribs (dull note on percussion) measuring 1 × 3 × 5 inches and weighing about 7 oz (80–300 gm). It lies in intimate contact with the undersurface of the diaphragm. This explains why a splenic abscess can rupture through the diaphragm causing empyema. When blood collects due to splenic injury, it irritates the diaphragm causing referred pain to the shoulder tip (**Kehr's sign**).

The **anterior border** is **notched** and is in contact with the stomach. It is enclosed by the **gastrocolic omentum**. The surgical importance of this is that when traction is applied on the stomach during vagotomy, a splenic capsular tear can occur, resulting in bleeding. The **two leaves** of gastrocolic omentum pass backwards in front of the kidney forming the **lienorenal ligament** (Fig. 26.2). This ligament is responsible for some part of the posterior fixation of the spleen. When this ligament is divided, the spleen can be brought to the surface of the wound during splenectomy.

The inferior part of the **hilum** of the spleen is (just) in contact with the **tail of the pancreas**. Often, during an emergency splenectomy, the tail of pancreas can get injured, resulting in a pancreatic fistula (Figs 26.2 and 26.3)

Splenic parenchyma

- It is **highly vascular**
- Splenic pulp has red pulp and white pulp.
- **Red pulp** is a honeycombed vascular space which is made up of cords of reticular cells and sinuses.
- **White pulp consists of lymphatic tissue** and lymphoid follicles containing lymphocytes, plasma cells and macrophages.

Blood supply

- **Arterial supply:** Splenic artery is a branch of coeliac artery. At the hilum of the spleen, it divides into 4–5 trabecular arteries and then branches into central arteries. Central arteries pass through white pulp and give radial branches to periphery including distal red pulp. Anatomical knowledge of these branches helps when performing partial splenectomy for ruptured spleen. (The blood flow rate through spleen is about 300 ml/min.)

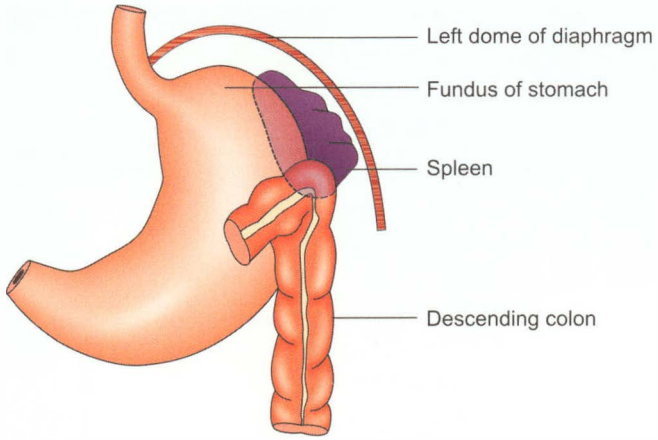


Fig. 26.1: Location of the spleen in relation to the fundus of the stomach and the diaphragm

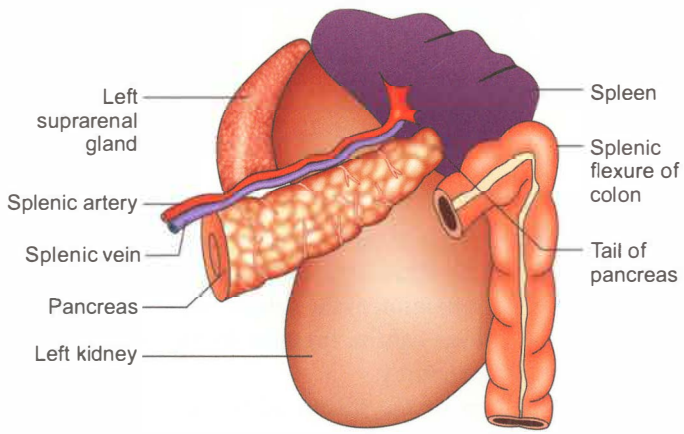


Fig. 26.2: Visceral relations of the spleen

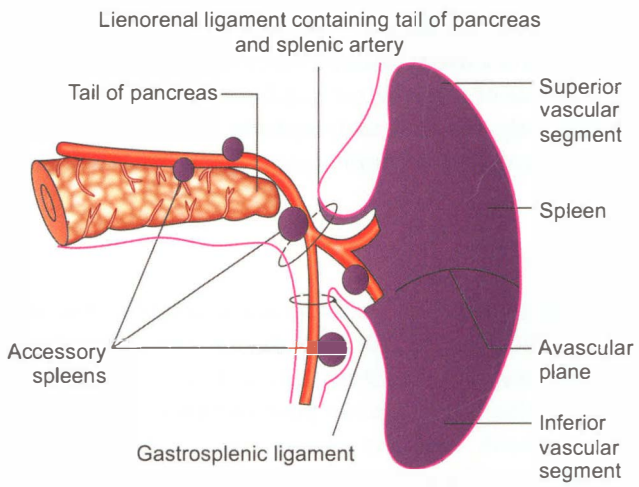


Fig. 26.3: Peritoneal ligaments attached to the spleen and common sites of accessory spleen

- **Venous drainage:** Four to five branches of the vein join and form the splenic vein. This forms the portal vein by joining with superior mesenteric vein behind the 2nd part of the duodenum.

Lymphatic drainage

The lymph drains into hilar nodes, then into retropancreatic nodes and finally into coeliac nodes. Since it has intimate contact with the fundus of the stomach and tail of the pancreas, spleen also used to be removed (nowadays not done) along with the stomach and tail of the pancreas during radical gastrectomy and distal pancreatectomy.

FUNCTIONS OF THE SPLEEN

1. **Destruction:** Removal of abnormal aged, damaged red blood cells by a process is called culling. Culling, the Greek word implies sorting a collection of two groups. One that will be kept and the other that will be rejected. For example, spherocytes, in hereditary, spherocytes are broken down in spleen. This happens in red pulp.
2. **Production:** Production of lymphocytes, monocytes and plasma cells. Lymphopoiesis continues throughout life. Lymphocytes take part in immune response of the body—largest.
3. **Antibody production:** Spleen synthesises antibody in its white pulp. It removes antibody-coated bacteria and antibody-coated blood cells via blood and lymph node circulation.
4. **Immune response:** White pulp is composed of malpighian corpuscles. They are composed of B lymphocytes and T lymphocytes—which are in periarteriolar lymphoid sheaths (PALS). Thus white pulp plays a major role in active immune response through humoral and all mediated pathways.
5. **Reservoir of platelets:** It is a storehouse of platelets, which are released into circulation in an emergency situation. Thus when spleen is enlarged, 80% of the platelets are sequestrated resulting in thrombocytopenia.

PEARLS OF WISDOM

Splenomegaly typically involves expansion of red pulp.

6. **Immune function:** It is the major site for synthesis of tuftsin, a peptide that stimulates phagocytic activity of leucocytes. Opsonins are antibodies produced by spleen against bacteria and fungi.
7. **Removal:** Heinz bodies, Howell-Jolly bodies, etc. are removed from RBCs. These cells are non-deformable intracellular substances.
8. **Erythrocyte production:** During foetal development. White and red blood cells production occur till 5th month of gestation. In adults in case of myelofibrosis erythopoiesis can occur.

CONGENITAL ABNORMALITIES

1. **Accessory spleens: Splenunculi:** They are found in about 10–20% of the population.
   - They can be found near the hilum of spleen, tail of pancreas, splenic ligaments or in the mesocolon.
   - In diseases of the spleen (haemolytic anaemia, thrombocytopaenic purpura), all these accessory spleens have to be removed along with spleen.
2. Splenic **agenesis** is rare.
3. **Polysplenia** is rare. It occurs due to failure of splenic fusion.
4. **Wandering spleen** is due to loose ligaments. It is more vulnerable for torsion.
5. **Hamartoma:** Rare, small masses of 1 cm in diameter. It may in lymphoid tissue also (Hamartoma = misfire).

RUPTURE OF THE SPLEEN

CAUSES

- **Blunt injury abdomen**
  - Injury to the left side of chest, left lower rib fractures, due to fall from a tree, road traffic accidents can be associated with splenic rupture.
  - Retroperitoneal haemorrhage, fracture spine and renal injuries may be associated with splenic injury.
- **Penetrating injuries** to the abdomen may cause **rupture of spleen**.
- **Spontaneous** rupture of the spleen is seen in malaria and infectious mononucleosis, rarely in sarcoidosis, haemolytic anaemia and leukaemia.
- **Iatrogenic:** Splenic capsule may be torn during surgical procedures such as vagotomy or gastrectomy due to traction on the stomach.

TYPES OF INJURIES

Most of these injuries are diagnosed by CT scan today (CT classification is given).

1. **Subcapsular haematoma:** A capsular tear or parenchymal tear results in subcapsular haematoma.
2. **Clean (incised wounds):** This can happen in sharp injuries, staal injuries (not common) or operative injuries.
3. **Lacerated wounds:** Often they are multiple as in blunt injuries abdomen.
4. **Hilar injuries** with **pedicle avulsion** are the most serious injuries. Need urgent laparotomy and ligation of bleeders and splenectomy. Otherwise, death will occur.
5. **Splenic injury** with other organs. Tail of pancreas, diaphragm, left kidney, left colon.

Splenic injury CT scan grading (Figs 26.4 to 26.6)

Grade I: Nonexpanding subcapsular haematoma <10% surface area
Nonbleeding capsular laceration with < 2 cm deep parenchymal involvement.

Grade II: Nonexpanding subcapsular haematoma 10–50% surface area.
Nonexpanding intraparenchymal haematoma <2 cm diameter
Bleeding capsular tear or parenchymal laceration 1–3 cm deep

Grade III: Expanding subcapsular haematoma or intraparenchymal.
Bleeding subcapsular. Haematoma or subcapsular haematoma >50% surface area
Intraparenchymal haematoma >2 cm
Parenchymal laceration >3 cm deep

Grade IV: Ruptured intraparenchymal haematoma with active bleeding.
Laceration involving segmental or renal vessel > 25% spleen volume devascularised

Grade V: Completely shattered or avulsed spleen.
Hilar laceration which devascularises entire spleen.

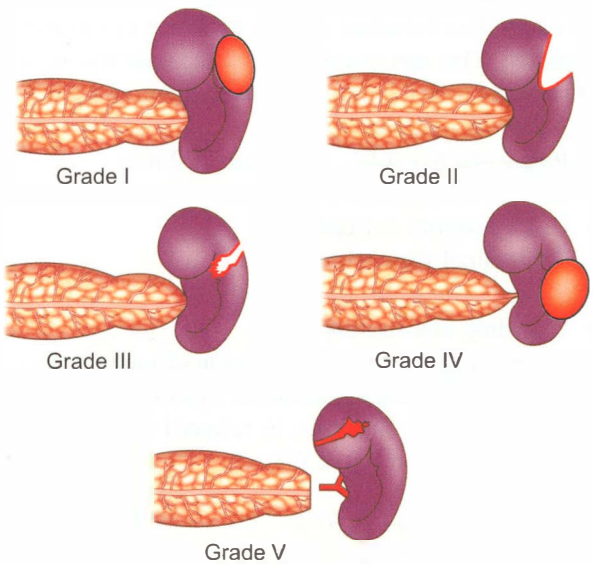


Fig. 26.4: Various grades of splenic injury

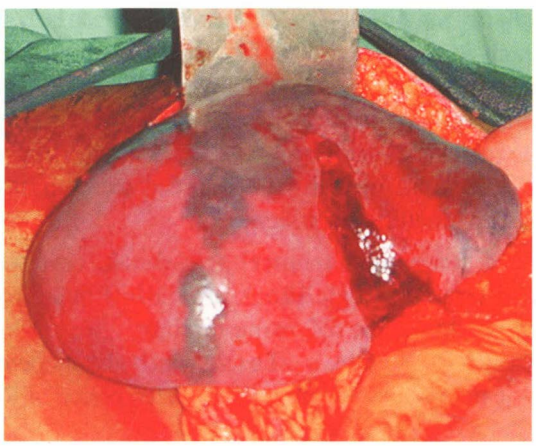


Fig. 26.5: Lacerated spleen

Fig. 26.6: Lacerated spleen—Grade III—splenectomy specimen

CLINICAL PRESENTATION

It can be divided into three groups:

1. **Tearing of splenic vessels:** Avulsion of splenic pedicle can result in severe haemorrhage and shock. Death can occur within a few minutes. Even in the best of the situations, death cannot be prevented often.
2. **A slow developing haemorrhagic shock** followed by recovery. Examination reveals evidence of intraperitoneal bleeding. This can occur due to a capsular tear or injury to the splenic parenchyma. Its clinical features are as follows:
   - Anaemia—pallor
   - Pulse—tachycardia, more than 100/min
   - BP—low/hypotension
   - Cold, clammy extremities
   - Abdominal distension
   - Paralytic ileus develops slowly
   - Guarding and rigidity
   - **Kehr's sign** is positive. Irritation of undersurface of the diaphragm by the blood causes irritation of phrenic nerve (C3, 4). Thus, this pain is referred to shoulder region (supraclavicular nerve C4).

PEARLS OF WISDOM

Kehr's sign can be elicited by elevation of the foot end of bed for about 10 minutes.

   - **Ballance's sign:** Blood in the vicinity of spleen is fresh blood which is coagulated and blood in the periphery is not coagulated. Hence, there will not be shifting dullness on the left side of the abdomen but can be present on the right side.
   - **Saegesser's splenic point of tenderness:** An area of tenderness on the left side between sternomastoid and scalenus medius (rare sign).
3. **Initial features of haemorrhagic shock, recovery and sudden haemorrhagic shock** after a few hours to a few days. It is due to the following reasons:
   - Greater omentum seals off a tear which gets reopened after some time. Time period is called "laten period of Randet".
   - A subcapsular haematoma which had developed ruptures after some time.
   - Associated injury to the tail of pancreas causes release of enzymes which digests the tissues at a later date. This variety is called "delayed type of shock" or delayed rupture of spleen.

PEARLS OF WISDOM

In polytrauma, if a hypotensive patient responds to intravenous fluids, it is a hypovolaemic shock. Head injury is rarely a cause of hypovolaemic shock.

COMPLICATIONS OF SPLENIC INJURIES

1. Haemorrhagic shock—if not treated—multi-organ failure results.
2. Disseminated intravascular coagulation due to massive bleeding and blood transfusion.
3. Injuries to the tail of pancreas—manifesting later as pancreatic ascites, pancreatic fistula or pseudocyst of pancreas.
4. Splenic artery pseudoaneurysm.
5. Splenic arteriovenous fistula.

INVESTIGATIONS

1. **Hb%** estimation, PCV (packed cell volume) estimation (however, in appropriate cases it should be repeated at frequent intervals to detect continuing haemorrhage).
2. **Four quadrant tap/aspiration** by using a fine needle (23 gauge) demonstrates blood either fresh or old. However, diagnostic peritoneal lavage **(DPL)** is more reliable (*vide infra*).
3. **Emergency USG** can reveal a splenic tear, a subcapsular haematoma and can rule out other injuries. It is the most important investigation in suspected cases of splenic injury.
4. CT scan is more reliable. It should be used in cases of doubtful diagnosis and stable patients.[1] It also rules out hollow viscus perforation, pancreatic injuries, etc (Fig. 26.7).

Fig. 26.7: CECT showing splenic and liver injuries

[1]Deaths have been reported in CT scan room, in unstable patients with haemoperitoneum.

5. Diagnostic peritoneal lavage (DPL)

- It is indicated in blunt injury abdomen where there are **equivocal signs** or **doubtful signs** of peritonitis.

Indications

- Unconscious patient with polytrauma with signs of abdominal injury.
- Unexplained shock
- Associated spinal cord injury

Procedure (Fig. 26.8)

- A 1 cm incision is made in the subumbilical region under local anaesthesia (LA) after emptying the bladder. The peritoneum is opened and a 12 or 14 Fr peritoneal dialysis catheter is introduced into the peritoneal cavity.
- The skin is closed and 1000 ml normal saline is allowed to flow over 30 minutes.
- The patient is turned to the right and left sides and fluid is allowed to flow out.
- **DPL is positive when**
  1. Fresh blood of more than 20 ml is removed immediately after inserting dialysis catheter and it does not clot.
  2. RBC count 1,00,000/mm^3 or WBC > 500/mm^3.
  3. Amylase level of over 175 units/dl.
  4. Bile, food or any other fresh contents.
  5. Gram stain is positive in the contents of lavage.
- Diagnostic accuracy is around 95%. DPL is useful to rule out intestinal injury in addition to intraperitoneal bleeding.

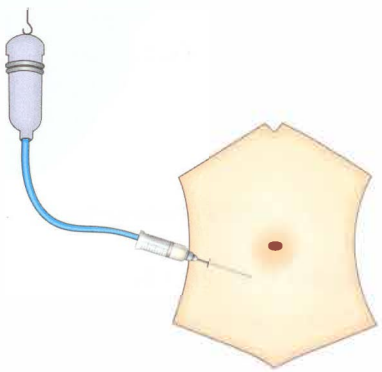


Fig. 26.8: Diagnostic peritoneal lavage

PEARLS OF WISDOM

DPL is more sensitive for mesenteric injury than CT scan. However in early diagnosis of hollow viscus injuries, DPL is superior to CT scan.

5. Plain X-ray abdomen[1] erect (**should not be done in an emergency situation**) when patients are in shock.

- Splenic outline may not be seen clearly.
- Fundic air bubble may be indented by the haematoma.
- Psoas shadow is obliterated (KUB X-ray).
- Evidence of fracture of lower ribs.
- Evidence of fluid in the peritoneal cavity—**'ground glass' appearance**
- It may detect free gas under the dome of diaphragm.
- Elevation of left dome of diaphragm.

TREATMENT (Key Box 26.1 and Fig. 26.9)

KEY BOX 26.1

TREATMENT—4 HEADINGS

- Emergency splenectomy – Massive bleeding and unstable
- Splenorrhaphy – Stable
- Partial splenectomy – Stable
- Nonoperative – Stable patient

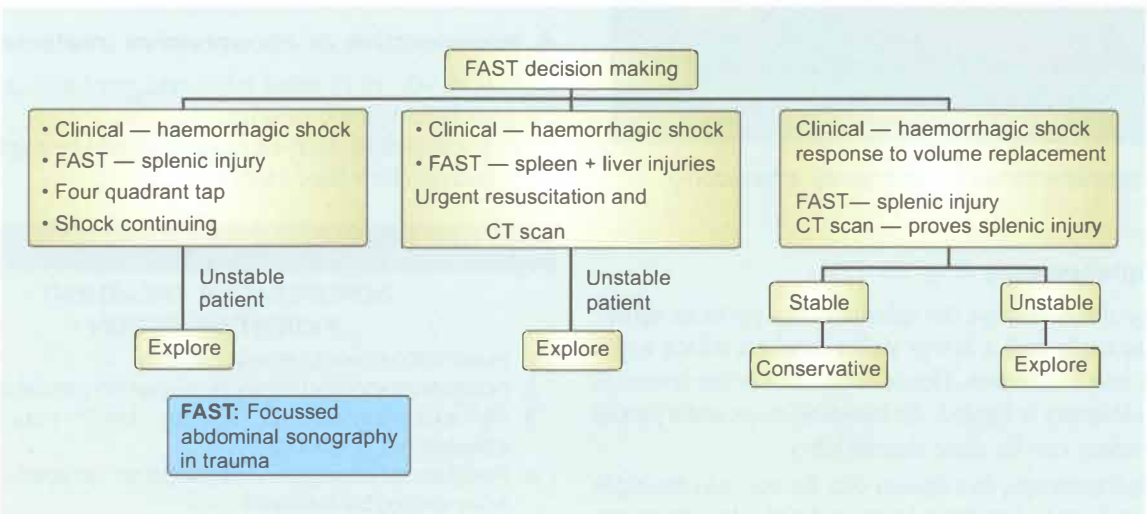


Fig. 26.9: Managment of splenic rupture

[1]Please do not make an unstable patient stand for the sake of X-ray. He may collapse due to hypotension.

1. Emergency splenectomy

When there is active bleeding with hypotension due to large lacerations, emergency splenectomy is the treatment of choice since it is quick, easy to perform and can be life-saving. The splenic artery is ligated first at the upper border of pancreas followed by splenic vein. In desperate situations, the spleen is mobilised by incising the lienorenal ligament, a large arterial clamp is applied at the splenic hilum and splenectomy is done (Figs 26.10 and 26.11).

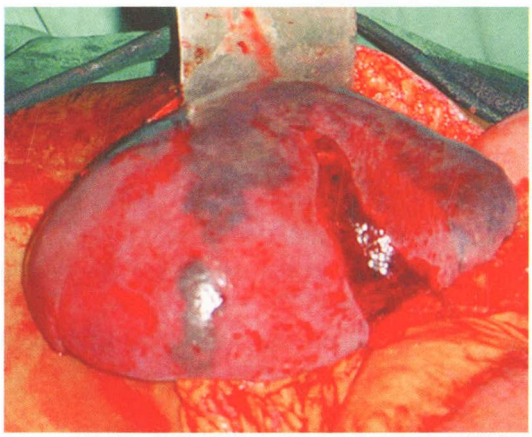


Fig. 26.10: Large laceration of the spleen

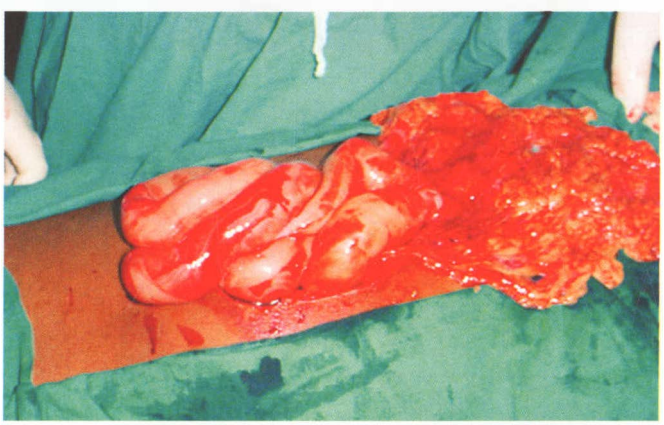


Fig. 26.11: Haemoperitoneum—emergency splenectomy

2. Partial splenectomy (Fig. 26.12)

- It can be done because the splenic artery gives an **upper polar branch and a lower polar branch** which again divides into 2 branches. Hence, when one of the branches of splenic artery is ligated, the bleeding stops and a partial splenectomy can be done comfortably.
- After splenectomy, the spleen can be cut into multiple pieces and can be implanted within the greater omentum. The spleen survives because of neovascularisation and functions like a spleen in the production of antibodies. This can be done in children.

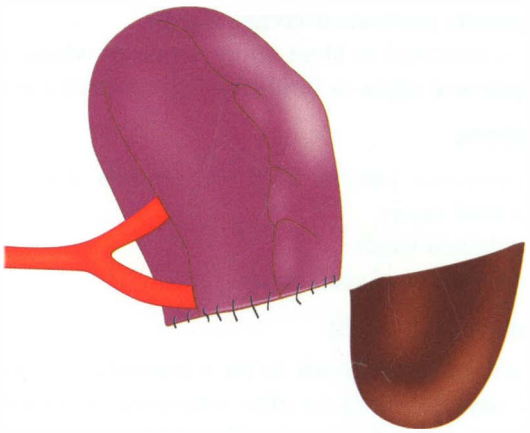


Fig. 26.12: Partial splenectomy

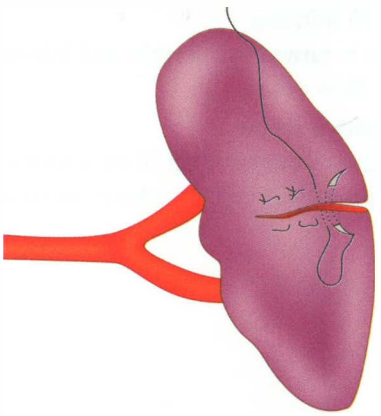


Fig. 26.13: Splenorrhaphy

3. Splenorrhaphy (Fig. 26.13)

- It is a method of preservation of spleen when the general condition of the patient is reasonably good.
- In this, a small tear is sutured by using chromic catgut and can be wrapped using greater omentum.

4. Nonoperative or conservative treatment

- It is safe in selected adult and paediatric patients with isolated splenic injuries.
- Small risk of delayed rupture should be explained to the patient (Key Box 26.2).

KEY BOX 26.2

NONOPERATIVE TREATMENT
PATIENT SELECTION

1. Haemodynamically stable patients
2. No other associated organ injuries which require laparotomy.
3. No extra-abdominal injuries which may preclude assessment of abdomen.
4. Facilities for emergency resuscitation, ultrasonography, CT scan should be available.
5. Patient must be willing for hospitalisation and close observation.
6. Regular monitoring by duty doctors, expert nursing staff who maintain hourly vital charts.

- Patients are admitted in intensive care unit. Serial monitoring of haemoglobin, total WBC counts are done. Abdominal girth and blood pressure are monitored. If there is any doubt of hypotension, exploration is done.

EARLY: COMPLICATIONS OF SPLENECTOMY

1. **Pancreatic injury (0–6%):** Treated by drain placement and measuring drain amylase. Usually it is a minor discharge, generally settles over a few days.
2. **Vascular injury (2–3%):** Slippage of ligature (splenic artery), hilar dissection, capsular tear during retraction. Needs to be identified and sutured or coagulated or clamped.
3. **Injury to bowel:** Colon, stomach. These can be avoided with careful dissection techniques. During ligation of short gastric arteries too close to the stomach side, injury to the stomach can occur. It is repaired in two layers. Colonic injury can occur during dissection to seperate the lower pole. It is also repaired in the similar manner.
4. **Diaphragmatic injury:** This can happen when adhesions are present between spleen and diaphragm due to perisplenitis or recurrent infarctions. Simple suturing is enough if recognised.
5. **Splenosis:** This refers to multiple small implants of splenic tissue on the peritoneal surface following traumatic rupture of spleen. They can give rise to adhesions.
6. **Pulmonary:** These are common complications. Generally get unnoticed. It can range from minor cough to atelectasis to pneumonia and pleural effusion. Basal atelectasis is one of the common causes of postoperative fever.
7. **Paralytic ileus:** Treated by drip and suction
8. **Haematemesis** can occur due to congestion of the gastric mucosa as a result of ligation of the short gastric vessels.
9. **Subphrenic abscess:** Not common. If it occurs, percutaneous drainage and antibiotics (Fig. 26.14).
10. **Wound problems:** Haematomas, seromas and wound infection.
11. **Thrombocytosis and thrombotic complications:** Sometimes it can be dangerous. They can present as abdominal pain and vomiting which is confused for incision pain, gastritis, etc. Ultrasound examination is a must. One of our patients had portal vein thrombosis which was managed by anticoagulants. Platelet coun after splenectomy was 8 lakh/mm^3.

Fig. 26.14: Post-splenectomy large collection

INDICATIONS FOR SPLENECTOMY

I. Always indicated
(a) Primary splenic tumour
(b) Hereditary spherocytosis

II. Usually indicated
(a) Primary hypersplenism
(b) Chronic ITP
(c) Splenic vein thrombosis
(d) Splenic abscess

III. Sometimes indicated
(a) Splenic injury
(b) Autoimmune haemolytic disease
(c) Elliptocytosis haemolysis
(d) Nonspherocytic congenital haemolytic anaemia
(e) Hodgkin's disease with anaemia
(f) Thrombotic thrombocytopenic purpura
(g) Idiopathic myelofibrosis
(h) Splenic artery aneurysm
(i) Wiskott-Aldrich syndrome (Key Box 26.3)
(j) Gaucher's disease

KEY BOX 26.3

WISKOTT-ALDRICH SYNDROME

- X-linked disorder predominantly presents as features of thrombocytopaenia such as epistaxis, bloody diarrhoea, petechiae in young age.
- Combined 'B' and 'T' cell immunodeficiency causes eczema
- More incidence of malignancies
- Spleen destroys platelets counts as low– 20,000 or 40,000/μL
- The best treatment is human leukocytic antigen (HLA) matched sibling bone marrow transplantation.
- Otherwise—splenectomy and antibiotics are the treatment of choice.

IV. Rarely indicated
- Chronic leukaemia
- Thalassaemia major
- Sickle cell anaemia
- Felty's syndrome
- Hairy cell leukaemia

HAEMATOLOGICAL INDICATIONS FOR SPLENECTOMY

IDIOPATHIC THROMBOCYTOPAENIC PURPURA (ITP)

- This condition occurs due to development of autoantibodies against patient's own platelets (Key Box 26.4).
- Normal levels of platelets are between 1,50,000 and 4,00,000 cells per mm^3.

KEY BOX 26.4

OTHER CAUSES OF PURPURA

- ↓ Production: Cytotoxic drugs, aplastic anaemia.
- ↑ Consumption of platelets: DIC as in septicaemia.
- ↑ Destruction: SLE, infectious mononucleosis.
- ↑ Capillary fragility: Steroid induced or Henoch-Schönlein purpura.

- The spleen is probably responsible for sequestration of platelets and for production of antibodies.

Types

Type 1: Acute—children: Follows an acute infection and resolves spontaneously in about two months.

Type 2: Chronic—adults: Longer than six months. No cause is identified.

Clinical features

- Condition is common in females, F : M :: 3 : 1.
- **Purpuric patches** (ecchymosis refers to skin discolouration due to extravasation of blood) are found on the buttocks and petechial haemorrhages (spots) are found on the limbs. These are **dependent areas** having high intravascular pressure (Figs 26.15 to 26.17).
- All types of haemorrhages are common and can be mild or moderate (Key Box 26.5).

KEY BOX 26.5

COMMON TYPES OF HAEMORRHAGES

- Epistaxis
- Menorrhagia
- Haematemesis
- Bleeding gums
- Haemarthrosis
- Haematuria

- Intracranial haemorrhage is found in 1–2% of patients but it may be the cause of death.
- In 25% of cases, the spleen is just palpable (small size).
- **Hess's tourniquet test:** More than 20 petechiae in a circle of 3 cm diameter in cubital fossa suggests purpura.

PEARLS OF WISDOM

If the spleen is big, it is not ITP.

Investigations

- Bleeding time is prolonged in purpura. Clotting and prothrombin times are normal.
- Platelet count is reduced.
- Bone marrow biopsy: Precursor of platelets (megakaryocytes) are increased.

Treatment

In children

- Spontaneous regression occurs in majority of cases.
- Short course of corticosteroids is beneficial. Tablet prednisolone in the dose of 10 mg/day over a period of 6 weeks is given.

In adults

Splenectomy is indicated in the following situations:

- ***When ITP has presented for more than 6–9 months.***
- ***When ITP has relapsed in spite of steroids,*** the patient is given a trial of prednisolone, 1 mg/kg/day. Platelet count rises within 7 days of starting steroids after which splenectomy can be done (Key Box 26.6).
- Also indicated if a patient has two relapses on steroid therapy or if the platelet count is less.

Fig. 26.15: Purpuric spots in the forearm

Fig. 26.16: Purpuric spots on the abdomen

Fig. 26.17: Severe sepsis with thrombocytopaenia

KEY BOX 26.6

SURGERY IN ITP

- Can be done even when platelets are as low as 10,000 cells/mm^3.
- **Diathermy** is used to open layers of the abdomen.
- **Ligate splenic artery first** at the upper border of pancreas after opening lesser sac.
- **Splenectomy is not difficult. Accessory spleens** (15–30%) if present, should be **removed**.
- Platelet transfusion may be required postoperatively, if there is bleeding.
- **Two-thirds** of patients will be **cured** by surgery.
- 15% will show improvement.
- Remaining will not benefit.

Results

70–80% of patients respond permanently and do not require any further treatment. Counts rise above 1,00,000/mm^3 in 7 days. Even if counts are not raised, recurrent bleeding rarely occurs.

Haemolytic anaemia

This type of anaemia results from an increased red cell destruction which occurs in the spleen. The life-span of red blood cells is also shortened. Hence, irrespective of the cause of haemolytic anaemia, these cases can benefit from splenectomy. **Anaemia, jaundice** and **splenomegaly** are the triad of haemolytic anaemia.

Causes of haemolytic anaemia

1. Hereditary spherocytosis
2. Acquired autoimmune haemolytic anaemia
3. Thalassaemia
4. Sickle cell disease

HEREDITARY SPHEROCYTOSIS

This hereditary disorder is transmitted by either parent as Mendelian autosomal dominant. The disease is characterised by a **defect in cell wall protein of the red cell, namely spectrin,** which increases reflux of sodium into the cell. This causes the biconcave red cell to swell and become spherical. Hence, the name spherocytosis.

Consequences of spherocytes in circulation

1. The spherocyte is already weak. Adding on to this, there is a greater loss of membrane phospholipid which results in a delicate fragile spherocyte.
2. Because of the altered shape, **oxygen and energy requirements of RBCs increase which cannot be met by the spleen**. In the spleen, they are destroyed and release excessive haemoglobin which is converted into bilirubin.
   - The amount of bilirubin produced is increased and most of it is unconjugated. It gets attached to albumin as it is **lipid soluble** and therefore, cannot be excreted in the urine. Hence, this type of jaundice is also known as acholuric jaundice.

PEARLS OF WISDOM

Most of this bilirubin is excreted in the biliary tree resulting in pigment stones in the biliary tree.

Clinical features (Figs 26.18 to 26.21)

- Commonly seen in young children but can manifest in adults also.
- Seen equally in both sexes.
- History of recurrent attacks of jaundice can be elicited from childhood.
- Jaundice is mild, never deep, not associated with itching or bradycardia.
- **Pallor** is an important feature due to destruction of the red cells.
- **Acute haemolytic crisis:** It is precipitated by an acute infection or stress. Abdominal pain, nausea, vomiting, pyrexia, pallor are the features. The condition can be confused for an abdominal emergency. In severe cases, anaemia, thrombocytopenia, leukopaenia can occur.
- **Spleen is moderately enlarged.** The liver is also palpable in a few cases.
- In adults, **gall stone colic** and obstructive jaundice due to **CBD** stone can also complicate the disease process. Incidence of gall stone disease is around 50%. **It is common after the age of 10.**
- Leg ulcers may occur as a result of anaemia.

PEARLS OF WISDOM

Young, pale, jaundiced, fatigued child with gall stones—Think of hereditary spherocytosis.

Investigations

1. **Peripheral smear: Spherocytes** are present.
2. **Reticulocyte count is increased (15–25%):** These are immature red cells which are discharged by the bone marrow due to loss of red cells.
3. **Coombs' test is negative.**
   - Serum bilirubin is mildly elevated and most of it is unconjugated.

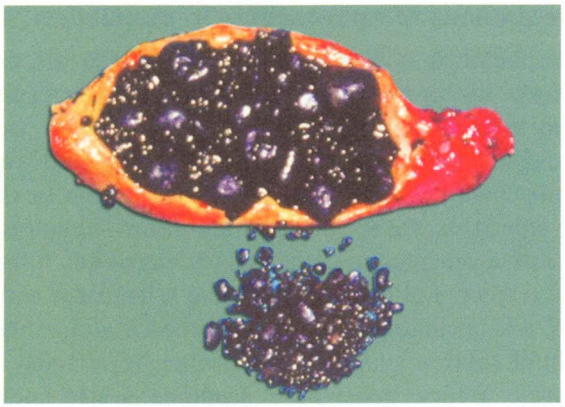


Fig. 26.18: Pigment stones

4. **Fragility test**
   - Normal red cell haemolysis occurs in **0.47%** saline solution. Here, it occurs at **0.6%** or even in weaker solution.
5. **Ultrasound** abdomen is done to rule out gall stones.

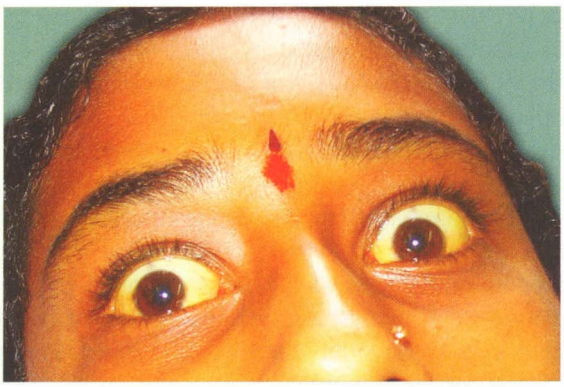


Fig. 26.19: Case of hereditary spherocytosis with faint jaundice in a 22-year-old girl

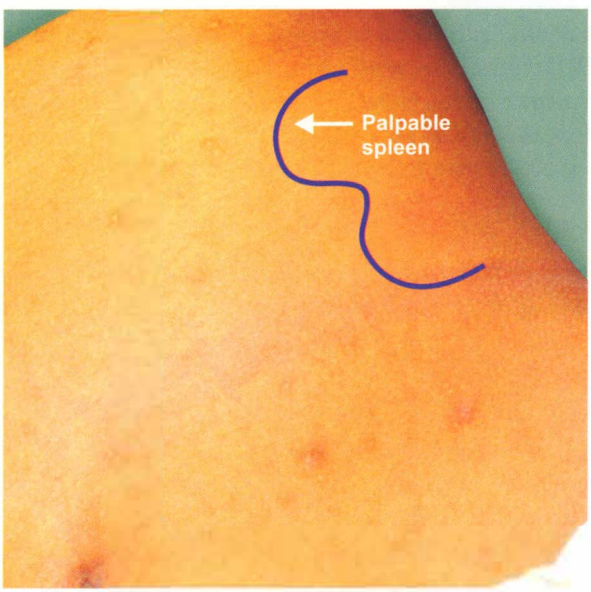


Fig. 26.21: Splenomegaly—its size was 5 cm

Treatment

Splenectomy is the treatment including removal of accessory spleen.

- Ideally, it should be done from the age of 6 to 10 years Why after 6 years? The incidence of post-splenectomy sepsis is more when splenectomy is done within 6 years of age. Why before 10 years? After 10 years of age, the incidence of gall stones increases.

Splenectomy helps in the following ways (Figs 26.19 to 26.22)

1. By decreasing erythrocyte destruction, anaemia improves, thus avoiding blood transfusions.
2. Erythrocytes achieve normal lifespan. Hence, jaundice also disappears.
3. Leg ulcers heal quickly.
   - If gall stones are also found in these cases, laparoscopic splenectomy and cholecystectomy can be done in the same sitting.

PEARLS OF WISDOM

Hereditary elliptocytosis is also a genetic disorder wherein RBCs are oval in shape. It also responds to splenectomy.

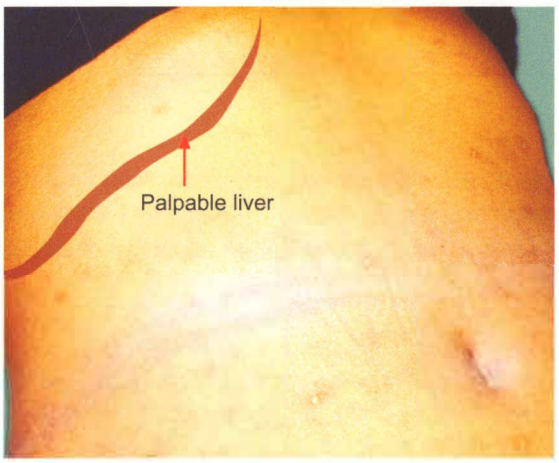


Fig. 26.20: She also had 2 cm enlarged liver and gall stones

Fig. 26.22: Ideal case—she underwent laparoscopic splenectomy and cholecystectomy

ACQUIRED AUTOIMMUNE HAEMOLYTIC ANAEMIA

Due to an autoimmune reaction, the red cell surface is damaged and it gets destroyed in the splenic red pulp. Antibodies develop against red cell antigens. In 50% of cases, the cause is not known. The following are the probable causes:

- SLE, lymphoma, chronic lymphocytic leukaemia
- Drugs: Methyldopa, mefenamic acid
- Exposure to chemicals or infections

Clinical features

- More common in females after 50 years of age.
- Splenomegaly, moderate in size
- Gall stones are found in around 25% of the cases.

Investigations

- Anaemia is present
- Spherocytes may be present because of damage to the red cell surface.
- Coombs' test is positive

Treatment

- It is a self-limiting disorder. Reassurance plays an important role.
- **Corticosteroids:** A short course of prednisolone 60 mg/day is given.
- Splenectomy is done when steroid response is not satisfactory. ***80% of patients respond to splenectomy***.

THALASSAEMIA

It is also called Cooley's anaemia, Mediterranean anaemia, Heinz body haemolytic anaemia.

Aetiopathogenesis

- It is a hereditary disorder, transmitted as a **dominant trait.** It is characterised by a defect in haemoglobin peptide chain synthesis. Depending on type of peptide chain involvement, it is classified into α, β and γ. However, β-thalassaemia is more common. There is a decrease in haemoglobin and red cells will be destroyed prematurely due to intracellular precipitates **(Heinz bodies)**.
- When it is a heterozygous disorder, it is called beta-thalassaemia minor and when homozygous, it is called beta-thalassaemia major (Cooley's anaemia). **The abnormal gene is inherited from either or both the parents.**

Clinical features

- Severe anaemia resulting in weakness, lethargy, leg ulcers, etc.
- Splenomegaly is mild to moderate in size. Sometimes, it is massive resulting in abdominal discomfort or may cause abdominal distension.
- Jaundice due to haemolysis. Liver can also be enlarged.
- To compensate for anaemia, bone marrow hyperplasia occurs resulting in bossing of skull and prominent malar bones. Thalassaemia major usually manifests in the first year of life.

Complications

1. **Haemosiderosis** (deposition of iron in tissues) of pancreas results in chronic pancreatitis and diabetes.
2. **Hepatic cirrhosis** due to liver haemosiderosis.
3. **Aplastic crisis** with severe life-threatening infections.
4. **Gall stones** occur in about 20% of cases.

Investigations

- Peripheral smear demonstrates microcytic hypochromic anaemia. Red cells are small, thin and are resistant to osmotic lysis.
- Haemoglobin electrophoresis reveals reduction or absent levels of haemoglobin A (HbA). There is a compensatory increase in foetal haemoglobin (HbF).

Treatment

- Blood transfusion: Multiple repeated transfusions may be necessary.
- Surgical: Splenectomy is indicated in a few cases who require multiple transfusions and patients with gross splenomegaly.

SICKLE CELL ANAEMIA OR SICKLE CELL DISEASE

This is a hereditary haemolytic disorder in which haemoglobin A is replaced by haemoglobin S (HbS). This results in crescent-shaped erythrocytes. This HbS molecule crystallises if there is hypoxia, strenuous work or due to dehydration. As a result of this, red cells are distorted and elongated. Therefore, blood viscosity is increased, causing obstruction in the splenic circulation (Key Box 26.7).

Effect of HbS blockage

- Splenic microinfarcts
- Splenomegaly or **autosplenectomy** due to repeated infarcts (Fig. 26.23).

Clinical features

- The disease is very common in Africans. Most of the patients in India have high HbF levels which protect HbS. Hence, symptoms are not seen in the first few weeks.

KEY BOX 26.7

FUNCTIONAL ABNORMALITIES IN SICKLE CELL ANAEMIA

- Spleen sometimes acts as a large reservoir for red cells and the sickle cells are destroyed.
- Antibody production is decreased.
- Spleen's ability to filter *Streptococcus pneumoniae* is reduced.

Table 26.1 Summary of haemolytic anaemias

	Aetiology	Shape of RBC	Spleen	Investigation	Treatment
1. Hereditary spherocytosis	Increase in red cell permeability to sodium	Spherocyte	Large	Reticulocyte count	Splenectomy very beneficial
2. Autoimmune haemolytic anaemia	SLE, drugs	Spherocyte	Medium size	Coomb's test is positive	Steroids: splenectomy in steroid failure cases
3. Thalassaemia (Cooley's anaemia)	Reduction of β-chain of haemoglobin	Small, thin and no shape	Moderate size	Resistance to osmotic lysis Electrophoresis HbA	Splenectomy for bulky spleen and in patients requiring frequent blood transfusion
4. Sickle cell	HbA is replaced by HbS	Sickle shape	Mild and later not palpable	Electrophoresis HbS	If hypersplenism +, splenectomy

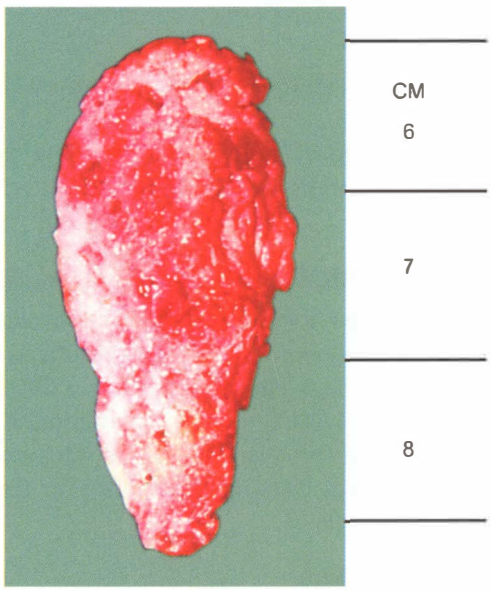


Fig. 26.23: Sickle cell anaemia—Nugget spleen (3 cm size). An example for autosplenectomy

- Abdominal pain is due to recurrent infarcts or due to gall stones in a few patients.
- Skin ulcers occur due to anaemia or hypoxia.

PEARLS OF WISDOM

Microinfarction of carpal and tarsal bones resulting in bony pains is described as hand-foot syndrome.

Investigations

- Peripheral smear demonstrates sickle cell.
- Haemoglobin electrophoresis to identify the type of haemoglobin.

Treatment

- Folic acid supplements
- Splenectomy has a doubtful value. It is indicated only when excessive numbers of RBCs are sequestrated causing anaemia (Table 26.1 for comparison).

Complications

- Infarct of cerebrum resulting in hemiplegia.
- Mesenteric infarction resulting in gangrene of the bowel.
- Pulmonary infarction resulting in chest pain.

SPLENECTOMY FOR OTHER CONDITIONS

1. Hypersplenism

It is defined as pancytopaenia in the presence of normal or hypercellular bone marrow. Following are the causes of hypersplenism:

- It usually involves red pulp expansion. It is to be remembered that in cirrhosis of the liver, 15% of patients develop hypersplenism. Anaemia and thrombocytopaenia are mild. Rarely requires splenectomy (a few causes are given below):

1. **Tropical splenomegaly** due to malaria, kala-azar or schistosomiasis, etc. Since these diseases are endemic in tropical countries, it is called tropical splenomegaly. Gross enlargement of the spleen and hypersplenism are indications for splenectomy in such cases.
2. **Myeloproliferative disorders:** Splenectomy reduces need for transfusion and may relieve abdominal discomfort.
3. **Portal hypertension:** Maximum benefit is in segmental portal hypertension with oesophagogastric varices (Fig. 26.24).
4. **Genetic:** Gaucher's diseases
5. Neoplastic infiltration
6. **Inflammatory disease:** Sarcoidosis, lupus erythematosus.
7. Chronic infections such as tuberculosis, brucellosis and malaria.

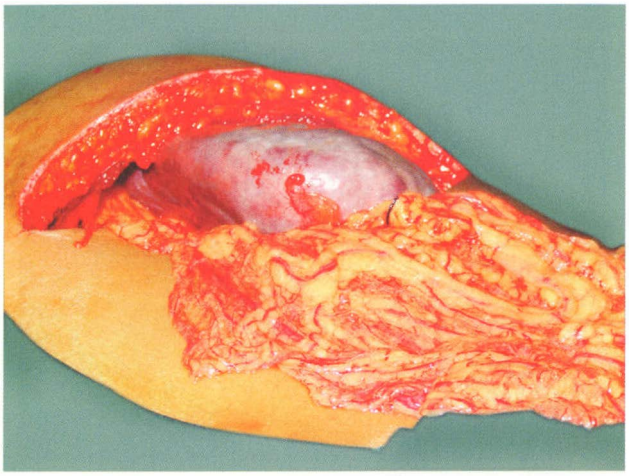


Fig. 26.24: Splenectomy done for left-sided portal hypertension due to splenic vein thrombosis

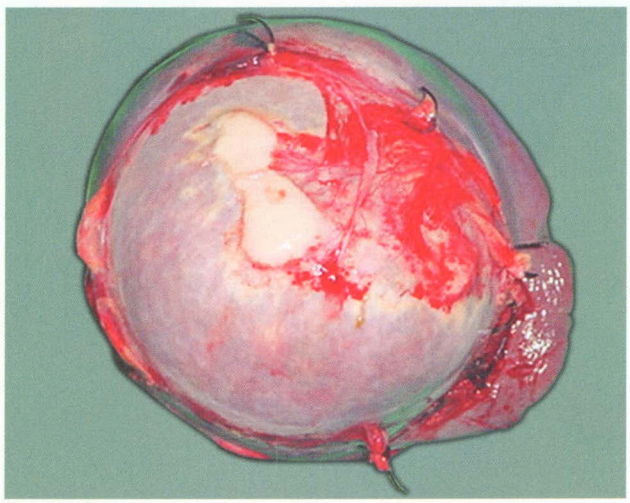


Fig. 26.25: Simple cyst of the spleen resulting in compression of the spleen—appearance like that of a ***globe***

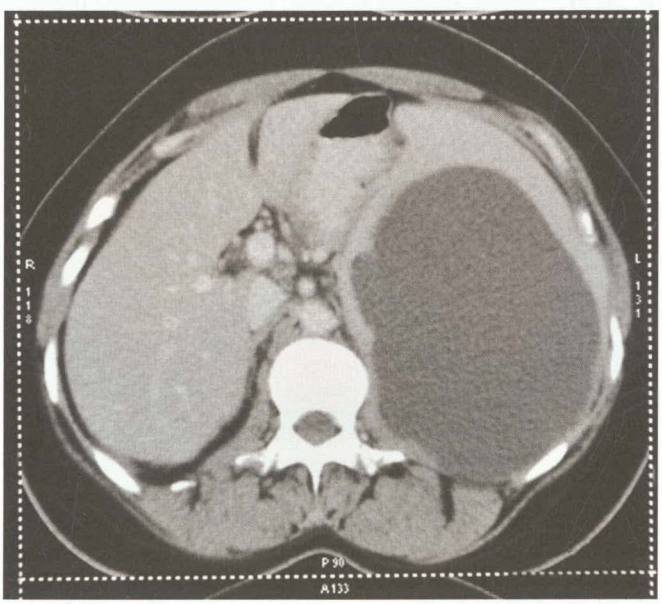


Fig. 26.26: Splenic cyst coronal view

8. **Chronic** haemolytic diseases—spherocytosis, thalassaemia, glucose-6-phosphate dehydrogenase deficiency, elliptocytosis.

2. Cysts of the spleen (Figs 26.25 and 26.26)

- ***Parasitic cyst*** is rare. Typically, it is caused by echinococcal disease **(hydatid disease)**.
- ***Traumatic cysts*** are due to a haematoma giving rise to liquefaction. These are false cysts of the spleen.
- ***Congenital cysts*** can be due to a dermoid cyst, haemangioma or lymphangioma.
- ***Better*** save the spleen by deroofing cyst wall.

3. Tumours of the spleen/neoplasms (Fig. 26.27)

- Among neoplasms, **lymphoma** is the commonest cause of ***enlargement*** of spleen. Spleen used to be removed as a part of a staging laparotomy. Now, it is very rarely removed. Patients with **large spleen** of chronic myeloid leukaemia, Gaucher's disease and hairy cell leukaemia (details later) will also benefit from splenectomy.
- **Fibrosarcoma** or **angiosarcoma** are rare malignant tumours.
- **Haemangioma** is the ***most common benign tumour*** of the spleen. It may turn into haemangiosarcoma.
- **Spleen** is removed in oesophagogastrectomy or upper carcinoma stomach—only when it is directly infiltrated. Sometimes it happens in carcinoma tail of pancreas/splenic carcinoma.

4. Other indications for splenectomy (Figs 26.28 to 26.31)

- **Splenic abscess** can occur due to infected septic emboli from otitis, typhoid fever or due to thrombosis of splenic vein causing infarction followed later by infection

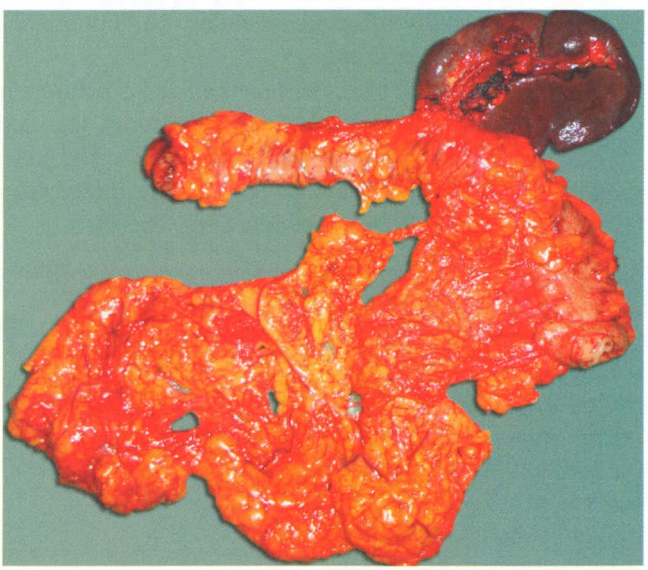


Fig. 26.27: Spleen removed with left hemicolectomy—advanced carcinoma

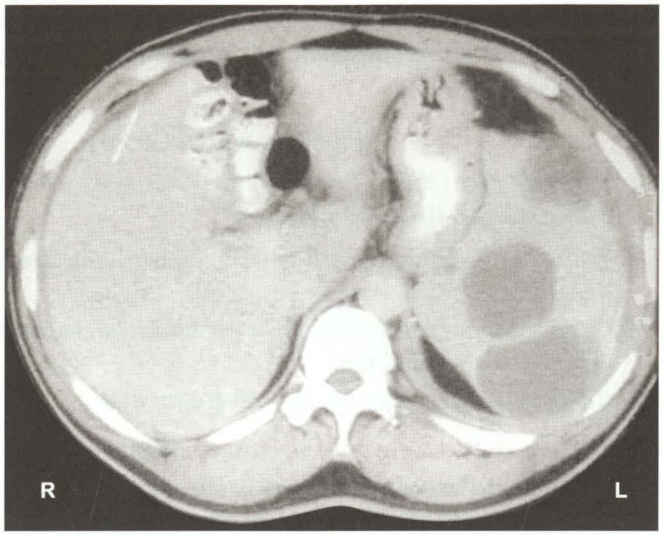


Fig. 26.28: CT scan showing multiple splenic abscesses

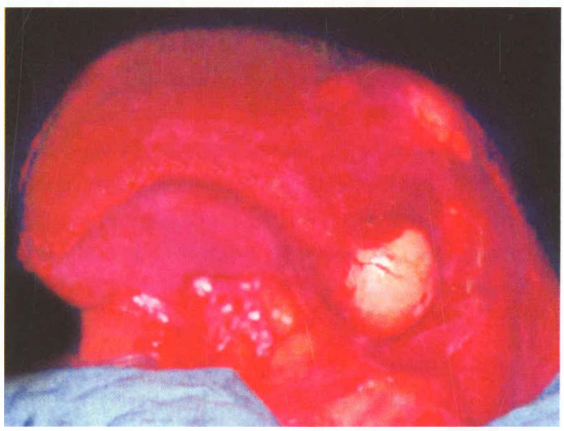


Fig. 26.29: Same patient as in Fig. 26.28 with multiple abscesses in the spleen

(Key Box 26.8). **Tubercular abscess** of the spleen is rare. It is usually affected secondary to abdominal or pulmonary tuberculosis.

- **Felty's syndrome** refers to splenomegaly, neutropenia with rheumatoid arthritis. After splenectomy, leg ulcers heal quickly and neutropenia also improves. The patient may respond to steroids better and the incidence of recurrent infection becomes less (Fig. 26.32).

KEY BOX 26.8

CAUSES OF SPLENIC ABSCESS

Salmonella infection
Paratyphi infection
Labour—puerperal sepsis
Ear infection—otitis
Necrosis of pancreas
Infection of bone—osteomyelitis
Chronic disease—tuberculosis

Remember as **SPLENIC**

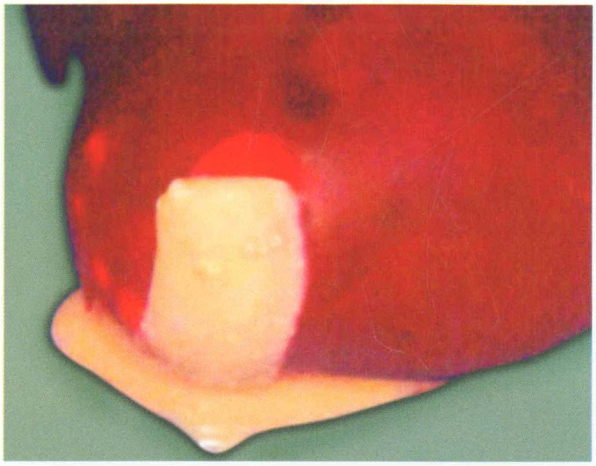


Fig. 26.30: Incision of the abscess after splenectomy

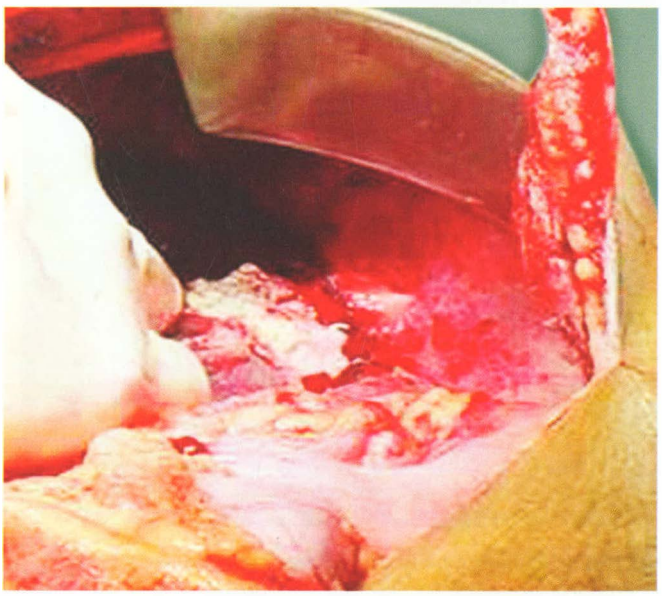


Fig. 26.31: Splenic abscess adhered to diaphragm and pre-renal fascia. In ITP, there will not be adhesions

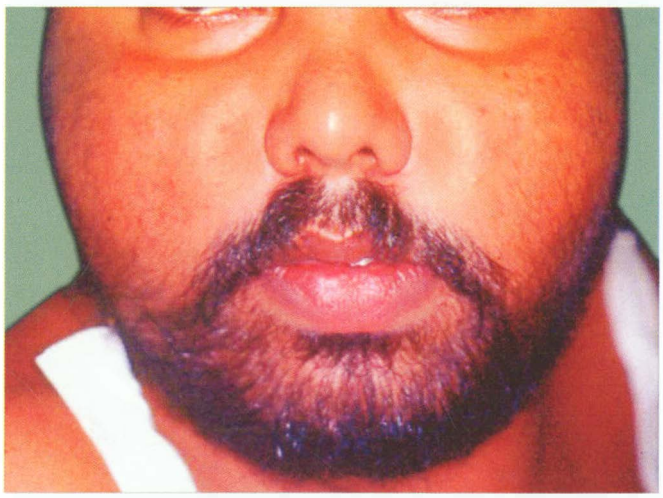
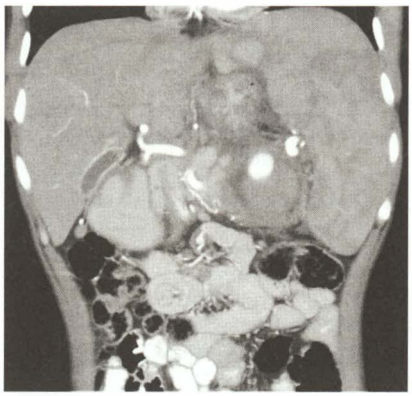
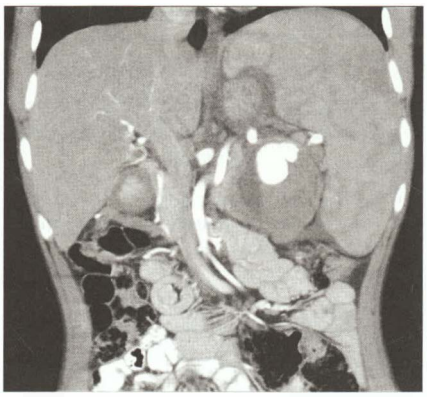
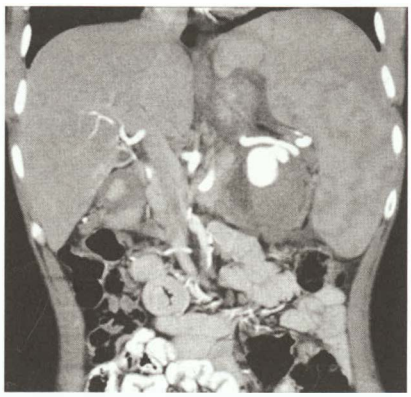


Fig. 26.32: Felty's syndrome. The patient had received steroids for one year. *See* the Cushingoid facies

Figs 26.33 to 26.35: A 28-year-old man presented with pain abdomen. CT scan showed splenic artery aneurysm with haematoma and thrombus suggestive of ruptured splenic artery aneurysm. He also had splenomegaly and features of portal hypertension. He was alcoholic. He might have had pancreatitis and developed splenic artery aneurysm (*Courtesy:* Prof Sampath Kumar, Dr Basavaraj Patil, Dr Dinesh, Dr Pawan Kumar Addala, Department of Surgery, KMC, Manipal)

5. Splenic artery aneurysm

It is an uncommon condition. More common in women, and some of them occur during *pregnancy*.

Causes

- **Atherosclerosis:** Commonly seen in elderly patients
- **Congenital:** Young patients. These patients may present for the first time during pregnancy with enlargement or rupture.
- **Acute pancreatitis:** Inflammatory process may give rise to pseudoaneurysm specially because of pancreatic necrosis.

Clinical features

- Pain and vomiting—haematemesis
- Thrill or bruit
- Calcification in a routine X-ray
- Rupture and features of haemorrhagic shock.

Investigations (Figs 26.33 to 26.35)

- Ultrasound/CT scan
- Angiography

Treatment

- **Nonoperative:** Embolisation of splenic artery.
- **Operative:** Ligation of splenic artery at the upper border of pancreas after opening lesser sac followed by splenectomy.
- **Rupture:** Carries high mortality rate.

6. Hairy cell leukaemia (HCL) (Fig. 26.36)

- The cells on a blood smear have an irregular outline due to the presence of filament-like cytoplasmic projections. Hence, the name **hairy cells**.
- HCL is a clonal proliferation of abnormal B cells (very rarely T cells).

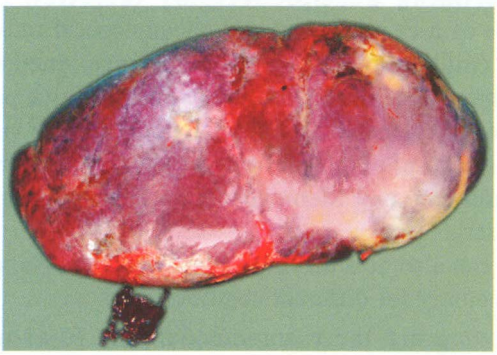


Fig. 26.36: Patient had pyrexia of unknown origin with pancytopaenia. Spleen was enlarged. Splenectomy was done and the biopsy report was non-Hodgkin's lymphoma

Clinical features

- Anaemia
- Recurrent infection
- Massive enlargement of spleen.

Investigations

- Blood counts are low. White cell count may be raised with circulating hairy cells.
- Bone marrow has increased cellularity with characteristic infiltration by **hairy cells**.

Treatment

- Drug: 2-Chloradenosine acetate (2-CDA) has been beneficial. It can induce remission.
- Splenectomy is indicated in cases of diagnostic difficulties and in very large spleens (Fig. 26.37).

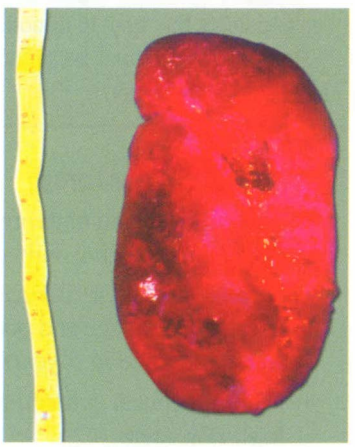


Fig. 26.37: Splenectomy was done for large spleen causing discomfort, reported as hairy cell leukaemia

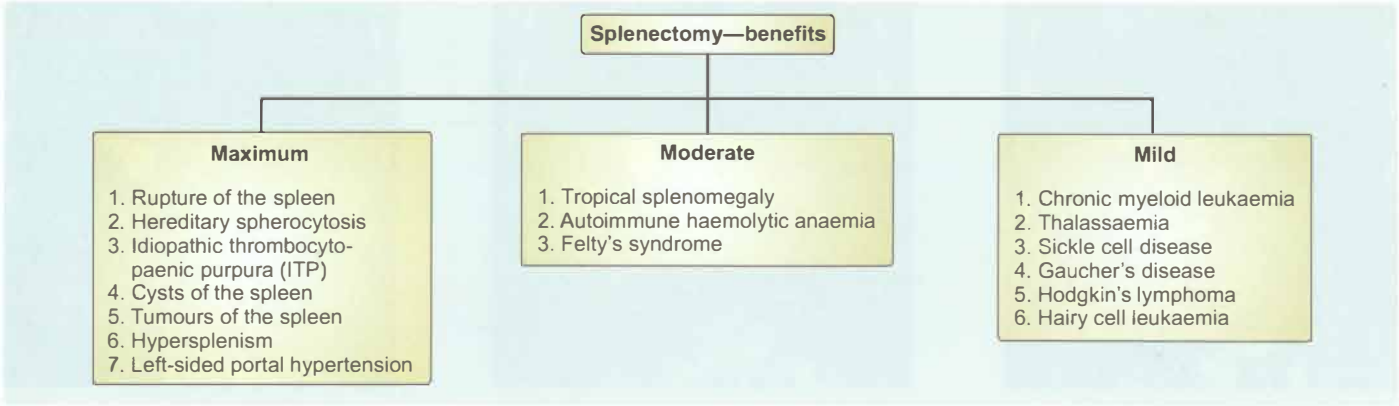


Fig. 26.38: Benefits of splenectomy

OPSI

Overwhelming postsplenectomy infection (OPSI): Incidence of meningococcal and pneumococcal infections is more in children after splenectomy. Hence, pneumococcal vaccine must be administered in the postoperative period.

- Following splenectomy, especially children are vulnerable for opportunistic infections such as Pneumococcus, *H. influenzae* and Meningococcus. Hence prophylaxis is necessary.
- OPSI can also be a major problem in adults who undergo splenectomy for different conditions.

Following are the recommendations; to avoid OPSI (Key Box 26.9).

KEY BOX 26.9

HOW TO AVOID OPSI?

- **O**ffer education—about spleen, role of splenectomy, side effects of splenectomy—especially in children.
- **P**rophylactic antibiotics before splenectomy.
- **S**peedy (prompt) treatment of infection. Early control of 'sepsis' is the 'key' to success.
- **I**mmunisation must be timely

Remember as **OPSI**

I. Elective splenectomy

- All children who undergo splenectomy before 5 years of age, should receive daily dose of penicillin until 10 years.
- *Haemophilus influenzae* type B vaccination for all irrespective of age and pneumococcal vaccine to be given (repeated once in 5 years).
- Meningococcal protection for only those who travel to high-risk areas.

PEARLS OF WISDOM

Vaccination should be given at least 2 weeks before elective splenectomy.

II. After emergency splenectomy

- Vaccines should be given as early as possible. However protection is **not always** guaranteed.
- Daily dose of oral prophylaxis with penicillin erythromycin or amoxycillin to all children till 10 years who undergo splenectomy before the age of 5.

Benefits of splenectomy (Fig. 26.38)

INTERESTING 'MOST COMMON' FOR SPLEEN

- Haemangioma is the most common benign tumour of the spleen.
- Lymphoma is the commonest cause of enlargement of spleen (in India, malaria is the cause).
- It is the most common solid organ injured in blunt abdominal trauma.
- Most common cause for splenic rupture is blunt abdominal trauma.
- Most significant benefit of doing a splenectomy for hypersplenism is from segmental left-sided portal hypertension.
- Most commonly performed investigation for suspected splenic trauma is abdominal ultrasound.
- Most common malignancy affecting spleen is lymphoma
- Most common complication of splenectomy is pulmonary complication
- Most common neoplasm of spleen—lymphoma
- Most common primary tumour of spleen—Haemangioma
- Most common primary malignant tumour of the spleen—haemangiosarcoma
- Most common infection after splenectomy is by *Streptococcus pneumoniae*
- Most common indication for splenectomy—splenic trauma
- Most common indication for splenectomy in elective setting—ITP.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- Splenic injury grades are added
- All topics have been updated
- New key boxes are added

MULTIPLE CHOICE QUESTIONS

1. In splenic injury, Kehr's sign refers to:

A. Absence of shifting dullness on the left side of abdomen

B. Tenderness on the left side between sternocleido-mastoid and scalenus medius muscle

C. Tender left iliac fossa

D. Shoulder pain due to irritation of the under-surface of the diaphragm

2. Spontaneous rupture of the spleen is seen in the following *except:*

A. Idiopathic thrombocytopenic purpura

B. Malaria

C. Infectious mononucleosis

D. Leukaemia

3. Diagnostic peritoneal lavage is said to be positive in all of the following *except*:

A. Fresh blood > 20 ml is aspirated after inserting dialysis catheter

B. Gram stain is positive in the contents of the lavage

C. RBC count > 100,000 cells/cu mm

D. Amylase level of 75 units/dl

4. The following is not true about nonoperative treatment in splenic trauma *except*:

A. Hemodynamically stable patient.

B. Hollow viscus injury

C. Close monitoring is available

D. Hospitalisation is required

5. Big spleen is not a feature of:

A. Malaria

B. Idiopathic thrombocytopaenic purpura

C. Portal hypertension

D. Leukaemia

6. The following is true about purpura:

A. Bleeding time is prolonged

B. Clotting time is prolonged

C. Prothrombin time is prolonged

D. Activated partial thromboplastin time is prolonged

7. Which of the following vaccination has to be given after splenectomy to prevent overwhelming post-splenectomy infection?

A. Measles vaccine

B. Pneumococcal vaccine

C. Hepatitis vaccine

D. Varicella vaccine

8. The following is true about splenectomy in idiopathic thrombocytopaenic purpura (ITP):

A. Is required in all cases of ITP

B. Must be done when platelet count is > 100,000 cells/cu mm

C. Accessory spleens, if present must be left behind

D. Two-thirds of patients will be cured by surgery

9. The following is true about splenectomy for hereditary spherocytosis:

A. Splenectomy should be done as a last resort

B. Splenectomy must be done either before 6 years or after ten years of age

C. Accessory spleen, if present must be left behind

D. Jaundice disappears after splenectomy

10. The most common benign tumour of the spleen is:

A. Haemangioma

B. Lymphoma

C. Tuberculoma

D. Lipoma

ANSWERS

1 D	2 A	3 D	4 B	5 B	6 A	7 B	8 D	9 D	10 A

27

Peritoneum, Peritoneal Cavity, Mesentery and Retroperitoneum

- Peritoneum
- Acute peritonitis
- Laparostomy
- Abdominal compartment syndrome
- Subphrenic abscess
- Special types of peritonitis
- Omentum
- Mesentery
- Misty mesentery
- Mesenteric cyst
- Retroperitoneum
- Retroperitoneal cyst, abscess, tumour
- What is new?/Recent advances

Introduction

Peritoneal cavity is the largest cavity in the body accommodating various viscera. It is divided into ***greater*** and ***lesser sac*** (omental bursa) which communicate through the ***foramen of Winslow*** or epiploic foramen. The peritoneum lining inner side of the parietes is called ***parietal peritoneum.*** It is ***very sensitive*** and is innervated by both somatic and visceral afferent nerves. This explains the ***sharp, localised, cutting*** pain of peritonitis. Diaphragm and central part of the peritoneum is supplied by phrenic nerve (C4) and partly by intercostal nerves. Rest of the peritoneum is supplied by intercostal nerves and lumbar nerves.

Lesser omentum: It is also called hepatoduodenal ligament. It extends from the duodenum to the liver. This has two layers and within these layers are the common bile duct, hepatic artery and hepatic portal vein.

THE PERITONEUM

Lining of peritoneum

The peritoneum is lined by a single layer of flattened cells and a thin layer of fibroelastic tissue. It is parietal peritoneum. A large peritoneal defect heals within a few hours because of these mesenchymal cells (flattened polyhedral cells—mesothelium). Applying this principle, some surgeons do not close the peritoneal layer after laparotomy. When parietal peritoneum is reflected into viscera, it is called visceral peritoneum.

It covers viscera and is supplied by autonomic nervous system. Hence, it is not sensitive. Thus, gastrojejunostomy can be done under local anaesthesia but distension and traction to the bowel causes pain. During herniorrhaphy under spinal anaesthesia, handling of bowel or traction on the bowel can produce uncomfortable upper abdominal pain.

Fluid

Peritoneal surface is a semipermeable membrane with an area comparable to that of cutaneous body surface. Nearly 1 m^2 of the total 1.7 m^2 area participates in fluid exchange with extracellular fluid space at the rate of 500 ml or more per hour.

It normally contains less than 50 ml fluid. When it is insulted by infection, a large amount of fluid can collect in this space giving rise to severe fluid and electrolyte imbalance. This is described as ***III space loss,*** e.g. ***peritonitis***, ***pancreatitis***. Peritoneal fluid helps in smooth gliding of intestines. Absorption of fluid and secretion of fluid are some important functions of peritoneum (Key Box 27.1).

KEY BOX 27.1

NORMAL PERITONEAL FLUID—TRANSUDATE

- Specific gravity below 1016
- Protein concentration less than 3 gm/dL
- White blood cell count less than 3000/μL
- Complement mediated bacterial activity
- Lack of fibrinogen related clot formation

Absorption and exudation

This takes place through capillaries and lymphatics present in between the two layers of peritoneum. This principle is applied in **dialysis**. The direction of circulation is towards subdiaphragmatic lymphatics.

Protective function

It secretes prostaglandins, interferons and free radicals which help in some protection against peritonitis.

ACUTE PERITONITIS

Definition: Inflammation of the peritoneum is called peritonitis.

Causes: They can be classified into **primary** or **secondary**.

I. Primary peritonitis

- Spontaneous peritonitis of childhood
- Spontaneous peritonitis of adults
- Tuberculous peritonitis
- Peritonitis associated with dialysis

II. Secondary peritonitis

This term refers to peritonitis from an intra-abdominal source and is the most common form of peritonitis. The following are the causes for secondary peritonitis (Fig. 27.1):

1. **Perforation of a hollow viscus**
   - Perforated duodenal ulcer, gastric ulcer
   - Perforated enteric ulcer, tubercular ulcer
   - Perforated Meckel's diverticulum
   - Perforated colonic ulcer
2. **Direct spread: Post-inflammatory**
   - Acute cholecystitis–gangrenous
   - Acute appendicitis
   - Gangrene of the intestine
   - Acute necrotising pancreatitis
3. **Penetrating injuries to the abdomen**, where the organisms gain entry from outside.
4. **Postoperative peritonitis** is due to the introduction of infection during surgery which might be due to:
   - Postoperative leaks
   - Foreign body (mop) in the abdomen
5. **Parturition peritonitis:** It refers to peritonitis after pregnancy and delivery.
6. **Blunt injuries to the abdomen**
   - Fluid which is spilled into the peritoneal cavity (example: Blood and bile can travel along paracolic gutter and manifest as pain in the right iliac fossa, causing guarding and rigidity. This has been called Valentino syndrome (*see* later in page 637).

PATHOGENESIS (Fig. 27.2 and Flowchart 27.1)

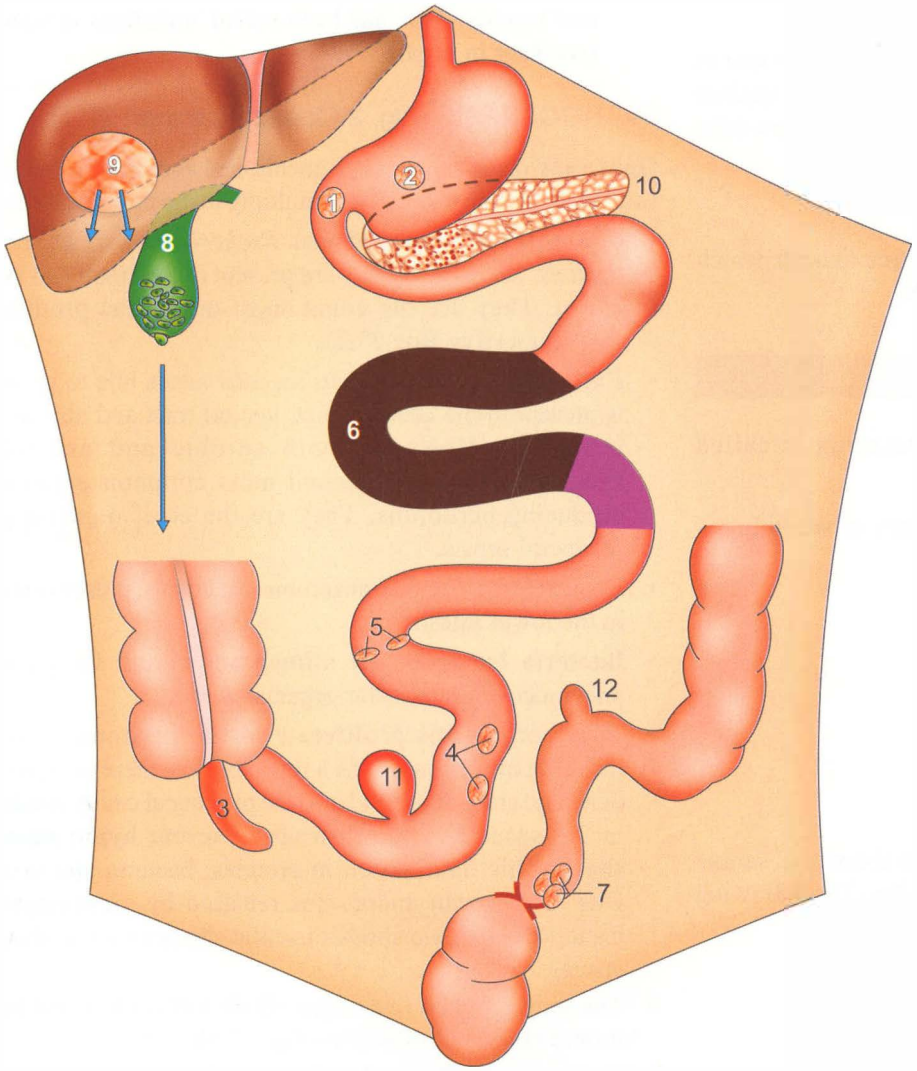
Due to any one of the reasons mentioned above, infection sets in and the causative organisms multiply in the peritoneal cavity.

- **Gram-negative organisms:** *Escherichia coli (E. coli), Proteus, Klebsiella*. They are present in the small and large bowel. They are the commonest organisms producing peritonitis (Key Box 27.2).
- ***Enterococci*:** *Streptococcus faecalis* needs bile to grow. It is present in the urinary tract, genital tract and also in the intestines. However, both aerobic and anaerobic streptococci are the second most common organisms producing peritonitis. They are the chief organisms in puerperal sepsis.
- **Bacteroides:** They are anaerobic organisms, present mainly in the lower intestine.
- **Bacteria from outside alimentary canal:** Gonococci, pneumococci, tubercular organisms, etc.
- These organisms proliferate in the peritoneal cavity resulting in peritonitis. As a result of this, there is secretion of a large amount of fluid into the peritoneal cavity resulting in 3rd space loss which leads to severe **hypovolaemic shock**. This fluid is rich in proteins, bacteria and toxins. Due to powerful endotoxins released by gram-negative bacteria, endotoxic shock or septic shock (*refer* to shock), ensues.
- The fluid is rich in fibrinogen which forms fibrin and helps in localisation of infection (Fig. 27.3).
- Peritoneum loses its shiny surface, becomes reddish and oedematous and is covered with thick fibrinous exudate.
- **Omentum:** It is a fatty apron with rich blood supply. A mobile double-layered peritoneal fold acts like a policeman to seal the area of infection or perforation. *Examples:* Perforated duodenal ulcer, acute appendicitis, acute diverticulitis, etc. Probably it also serves to supply collateral blood supply to the ischaemic viscera. It also has immunological functions such as supply of phagocytes which destroy unopsonised bacteria.

KEY BOX 27.2

ORGAN-SPECIFIC ORGANISMS

- **Gastric perforations:** Sterile with minimum gram-positive organisms.
- Ileal perforation, appendicitis—aerobic bacteria in 30% patients, anaerobic in 10% patients. Predominant aerobic bacteria include gram-negative *E.coli*, Streptococci, Proteus and Klebsiella.
- **Colonic-rectum:** Faecal spillage produces a load of 10^{12} or more—gram-negative and anaerobic bacteria per gram of stools.



Common causes of perforation
1. Perforated duodenal ulcer
2. Perforated gastric ulcer
3. Perforated appendicitis
4. Perforated enteric ulcers
5. Perforated tubercular ulcers
6. Gangrene intestine
7. Perforated carcinoma left colon

Uncommon causes of peritonitis
8. Perforated gall bladder
9. Ruptured amoebic liver abscess
10. Necrotising pancreatitis
11. Perforated Meckel's diverticulum
12. Sigmoid diverticular perforation

Fig. 27.1: Common causes of generalised peritonitis

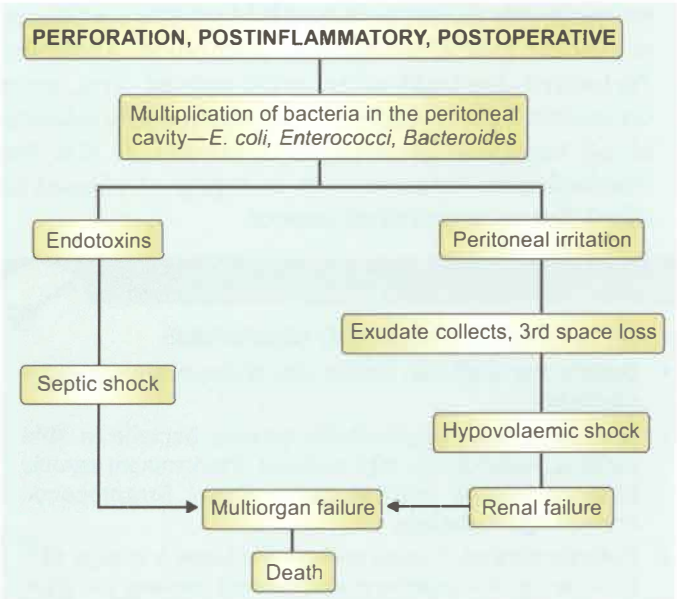
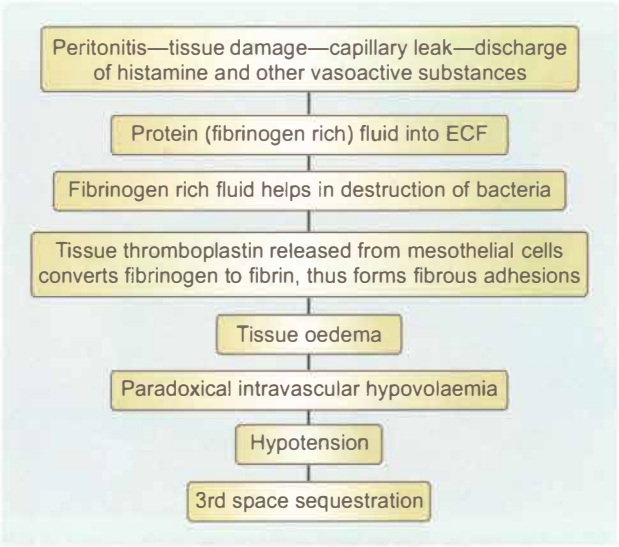
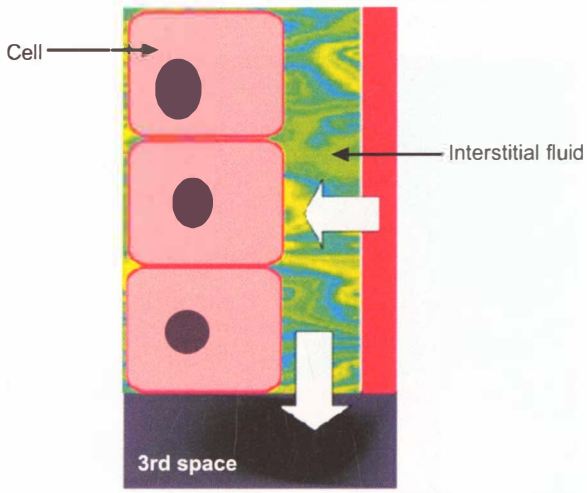


Fig. 27.2: Pathophysiology of peritonitis

Fig. 27.3: Fibrin plaques: Early cases of peritonitis with fibrin plaques all over the peritoneal cavity

Flowchart 27.1: Pathogenesis of peritonitis





Fluid sequestration

Types of peritonitis

A. Local peritonitis: If a perforation is small and if it is sealed off immediately by omentum, it will give rise to local peritonitis. Examples: Small gastric ulcer perforation or diverticular perforation, gall bladder perforation. Anatomical factors also play a role in local peritonitis. *Examples*: Retrocaecal appendicitis with perforation. It is behind the caecum and in retroperitonaeum. Signs are confined to right iliac fossa only. In posterior gastric perforations or acute pancreatitis, signs are limited to upper abdomen. Pelvic peritonitis is another example—occur following septic abortions or salpingo-oophoritis.

B. Generalised peritonitis: If the contents of the viscus leak into the peritoneal cavity with force, as it occurs in intestinal perforation or due to perforation of a free lying organ—example: Meckel's diverticular perforation. Virulence of bacteria is more as in colonic perforations with generalised peritonitis. Duodenal ulcer perforation can manifest as severe pain in the right iliac fossa mimicking appendicitis. Many have been operated as appendicitis. Reason being a broad right paracolic gutter and the contents travel down into right iliac fossa. This has been referred to as **Valentino syndrome** (Key Box 27.3).

KEY BOX 27.3

VALENTINO SYNDROME—VALENTINO APPENDIX

- **Rudolph Valentino** was an Italian actor who lived in early 20th century.
- On August 15, 1926, he was admitted with the diagnosis of appendicitis and gastric ulcers, with peritonitis.
- He underwent appendicectomy.
- Continued to have peritonitis
- He developed pleural effusion and sepsis
- Dies after a few days of surgery.
- In retrospect what he had was duodenal ulcer perforation.
- It is also called **Valentino syndrome** because in any inflammation of the upper abdominal viscera, contents can travel down the right paracolic gutter into the right iliac fossa resulting in pain and tenderness mimicking acute appendicitis.

FACTORS DECIDING THE SEVERITY OF PERITONITIS

- **Clean perforation:** Upper GI-Gastric juice remain sterile for 6–8 hours. Hence, in early stages, there will be mild chemical peritonitis and early treatment is give good results.
- **Distal gut perforation and infected bile peritonitis:** Very dangerous and severe, causing sepsis and septic shock early.
- Postoperative peritonitis that usually occurs due to anastomotic leak is also dangerous.
- A perforation sealed off early by omentum causes mild peritonitis. Retrocaecal appendicitis produces minimal local peritonitis
- On the other hand, perforated Meckel's diverticulitis produces diffuse peritonitis soon (Key Box 27.4).
- A few causes of peritonitis are shown in Figs 27.4 to 27.18B.

KEY BOX 27.4

FACTORS AFFECTING DIFFUSE PERITONITIS

- Speed of peritoneal contamination, e.g. perforation of Meckel's diverticulum
- Stimulation by purgatives
- Virulence of organisms
- Perforation in a closed loop obstruction
- Immunocompromised status
- Young children. Omentum is thin and small

Clinical features

It depends upon whether it is localised peritonitis or generalised. In cases of retrocaecal appendicitis, the abdominal signs may be minimal but guarding and rigidity of the back muscles is characteristic. Features of generalised peritonitis are as follows:

A FEW CAUSES OF PERITONITIS

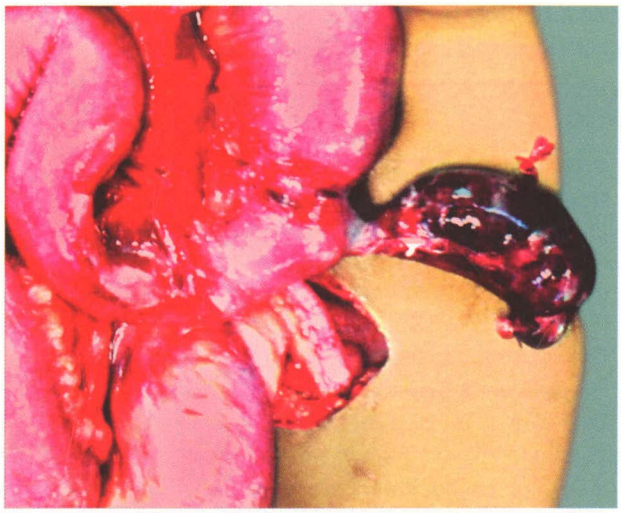


Fig. 27.4: Meckel's diverticulitis

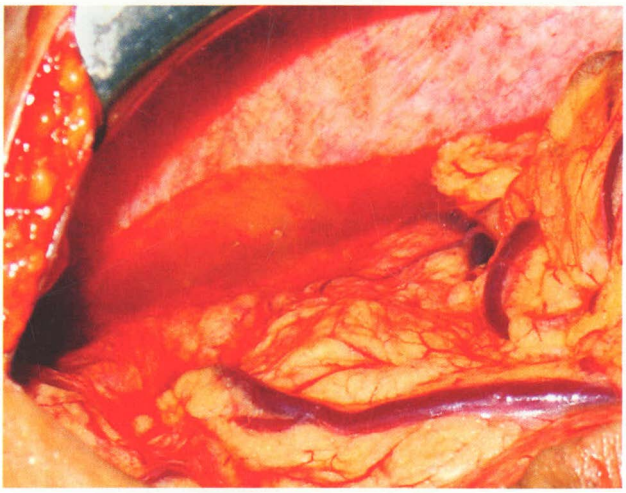


Fig. 27.5: Peritonitis due to ileal perforation consequent to tuberculosis. Usually it is an ulcerative variety

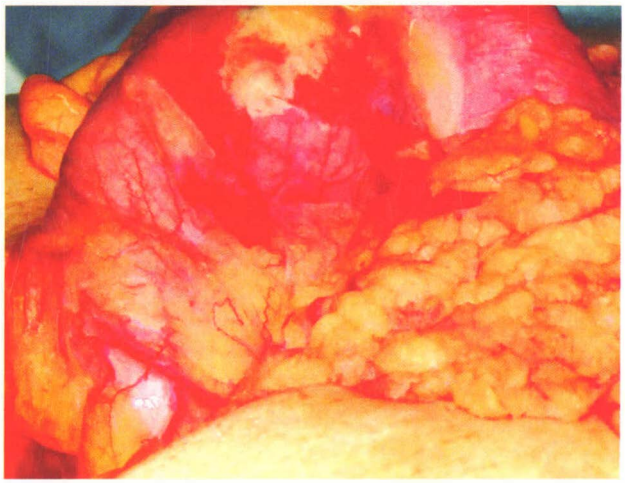


Fig. 27.6: Transverse colon injury due to steering wheel—simple closure in early cases without much peritonitis. Otherwise resection/closure with or without diversion ileostomy may be required

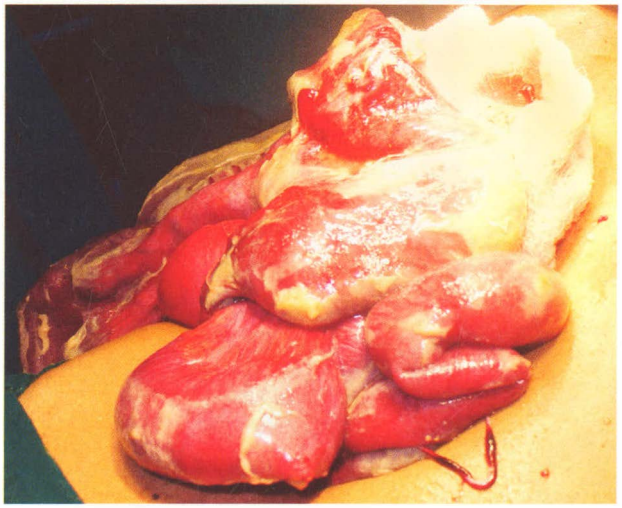


Fig. 27.7: Enteric perforation which is 4 days old—very friable edges. Re-leak after suturing is common

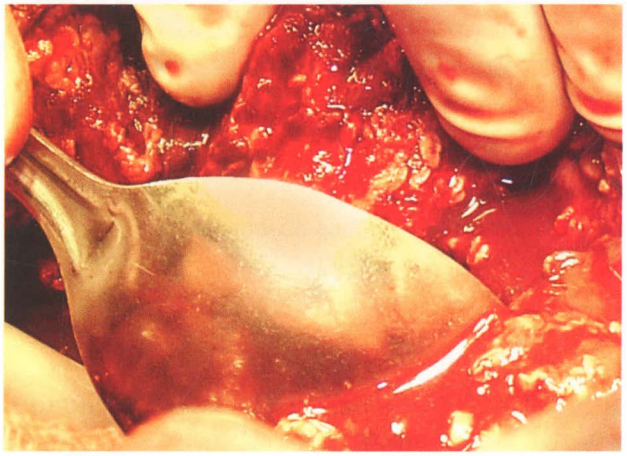


Fig. 27.8: Pancreatic necrosis—necrosectomy is done—you can see spoon has been used to remove necrotic tissue. Patient was in sepsis and peritonitis. Dramatic recovery took place

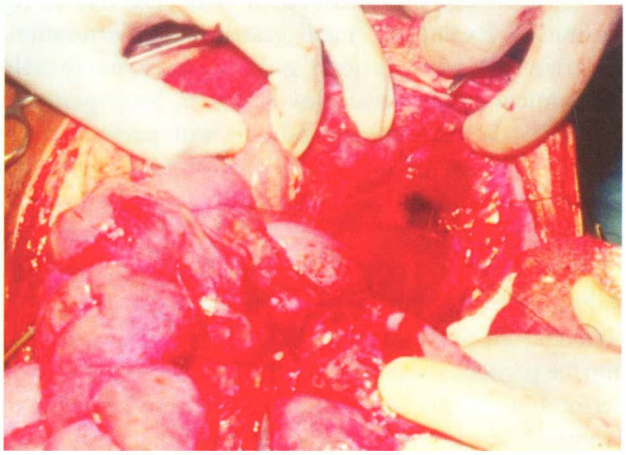


Fig. 27.9: Chronic duodenal ulcer perforation resulting in biliary peritonitis. Golden time to operate is within 6 hours before bacterial peritonitis sets in

A FEW CAUSES OF PERITONITIS

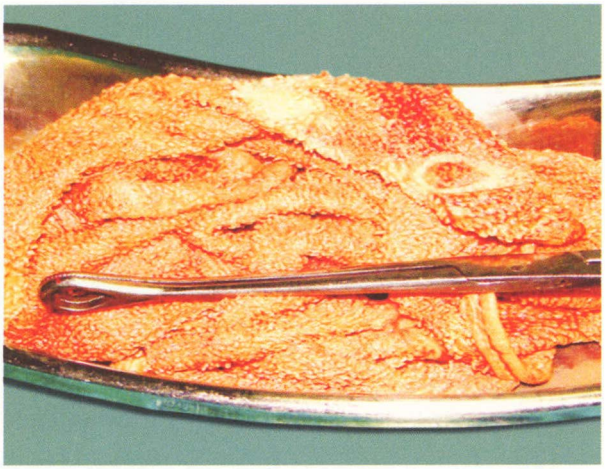


Fig. 27.10: Postoperative peritonitis due to a mop left behind following caesarean section. Always count the mops and instruments before closure of the abdomen. It is a good practice

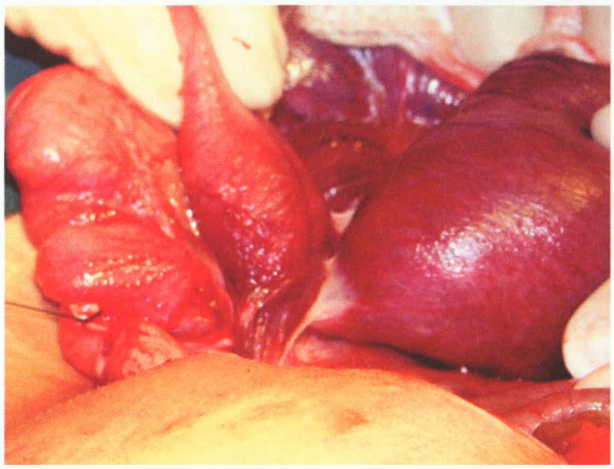


Fig. 27.11: Fibrous band causing gangrene of the terminal ileal loop resulting in peritonitis. Patient had pelvic peritonitis due to tuberculous salpingitis. That resulted in bands

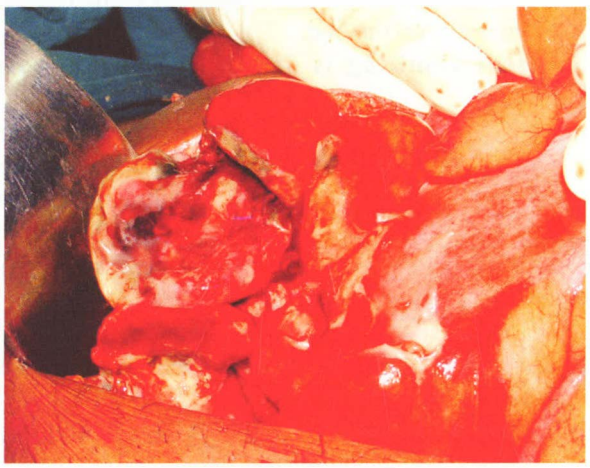


Fig. 27.12: Sigmoid diverticular perforation causing peritonitis. Sigmoid colon is the most common site of acquired diverticuli

Fig. 27.13: Faecal peritonitis and faecal fistula due to anastomotic leak following right hemicolectomy. Ischaemia and tension are the two common causes for the leak

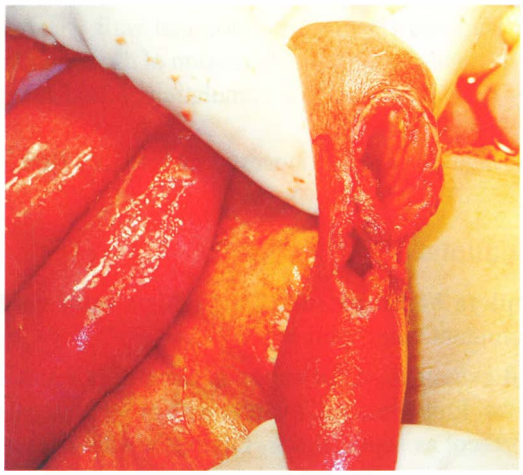


Fig. 27.14: Proximal jejunal transection following blunt abdominal trauma. It is one of the common sites affected in blunt abdominal trauma

Fig. 27.15: Colostomy gangrene —the intra-abdominal segment was also gangrenous. It is important to always check for the vascularity of the colostomy site before closing abdominal incision

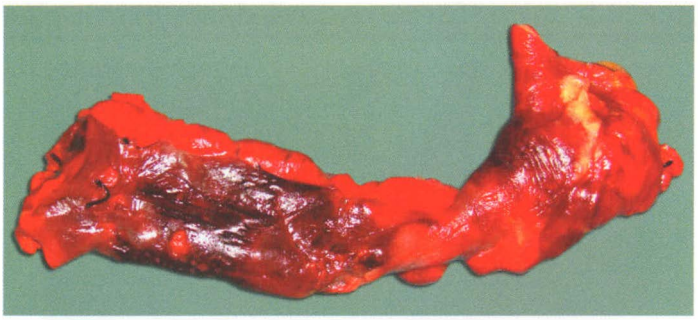


Fig. 27.16: Retracted colostomy—intra-abdominal gangrenous segment of the intestine causing peritonitis. It was resected

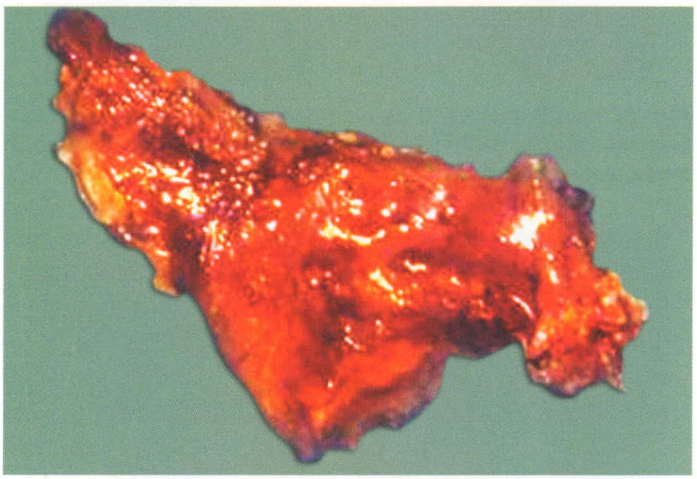


Fig. 27.17: Necrosectomy specimen—acute pancreatitis

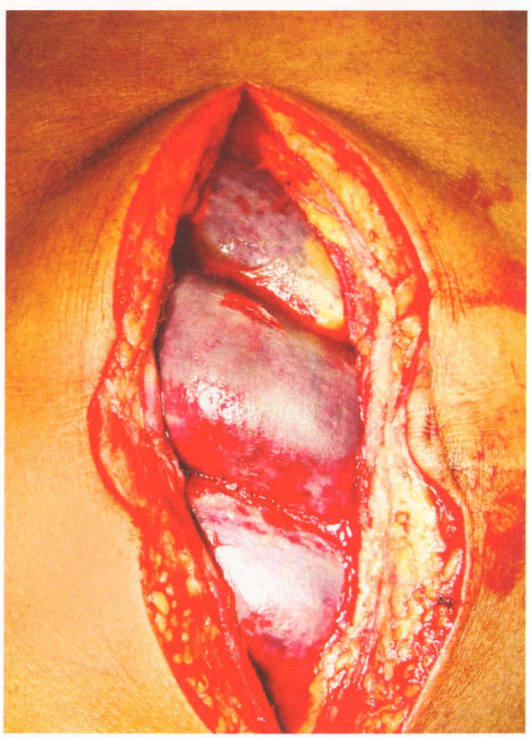


Fig. 27.18A: Mesenteric ischaemia giving rise to gangrene

Fig. 27.18B: Massive gangrene following superior mesenteric arterial occlusion—patient had severe acidosis

- **Severe abdominal pain** which is cutting in nature, becomes worse on movement of the abdominal wall. Hence, the patient lies still on the bed.
- **Persistent vomiting** is due to irritation of parietal peritoneum.
- **The pulse rate is increased.** An increase in the pulse rate may be an early indication of peritonitis, in cases of gangrene of the bowel or peritonitis following perforation of bowel.
- **High-grade fever with chills and rigors** indicates a septicaemic process.
- **Cough tenderness** indicates parietal peritoneal inflammation. Abdominal tenderness is elicited in all quadrants of the abdomen.
- **Rebound tenderness** (Blumberg's sign): Abdomen is pressed for a few seconds. The patient experiences pain. Sudden release of pressure causes severe pain. It is due to sudden movement of the sensitive parietal peritoneum (Fig. 27.19).
- **Guarding** and **rigidity** of abdominal wall.
- Bowel sounds are absent. Distension of the abdomen occurs within a few hours due to accumulation of fluid and paralytic ileus.
- **End-stage disease:** Hippocratic facies (Key Box 27.5 and Fig. 27.20).

Investigations

1. **Complete blood picture** shows **high total count** with predominant neutrophil count.
2. **Blood** examination for **sugar** is done to rule out diabetes mellitus. Empyema gall bladder with or without perforation can present as septic shock. Often, they are diabetic.
3. **Plain X-ray abdomen, chest and upright**
   - **Gas under the diaphragm**—perforation (Figs 27.21 to 27.23)

KEY BOX 27.5

HIPPOCRATIC FACIES

- Hollow, bright eyes
- Pale and pinched face
- Cold perspiration in the head and brows
- Blue lips
- Dry, cracked tongue

Fig. 27.19: Rebound tenderness is the diagnostic sign of peritonitis

Fig. 27.20: Hippocratic facies: Sunken eyes, drawn in cheeks, dehydrated, blood in the Ryle's tube

- **Ground glass appearance**—a smooth homogeneous appearance due to accumulation of fluid (Fig. 27.24).
- Air in the bowel wall—gangrene (Fig. 27.25)
- Obliteration of psoas shadow and preperitoneal fat planes.

4. Abdominal USG to detect **fluid in the abdomen**.

Following are different fluids which may give clue to the diagnosis

- Frank pus—peritonitis of more than 48 hours old

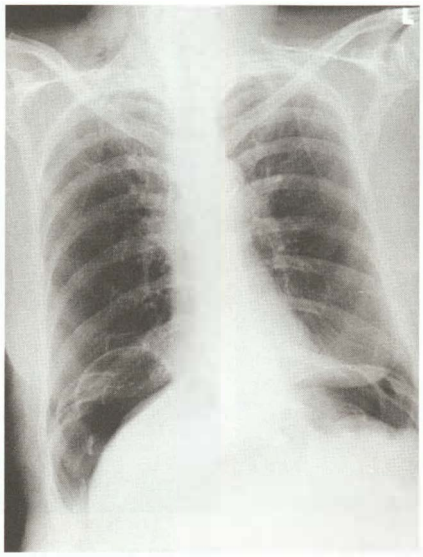


Fig. 27.21: Chest X-ray showing free gas under diaphragm

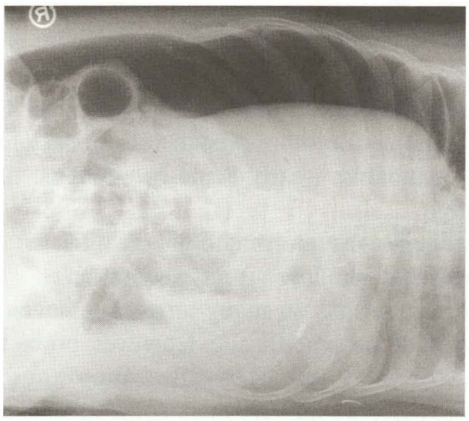


Fig. 27.22: Lateral decubitus X-ray showing free gas in the peritoneal cavity

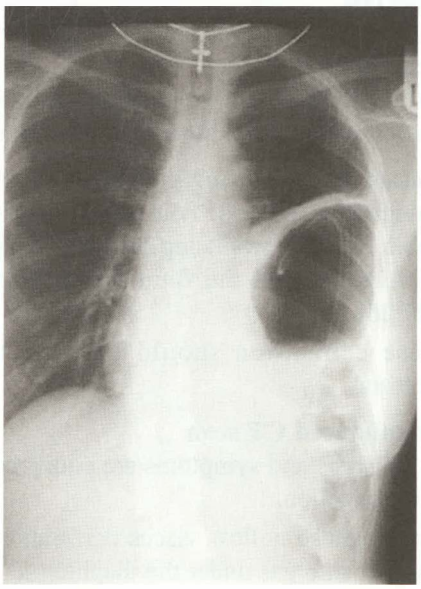


Fig. 27.23: Diaphragm is elevated—fundic air bubble is in the chest—traumatic diaphragmatic hernia

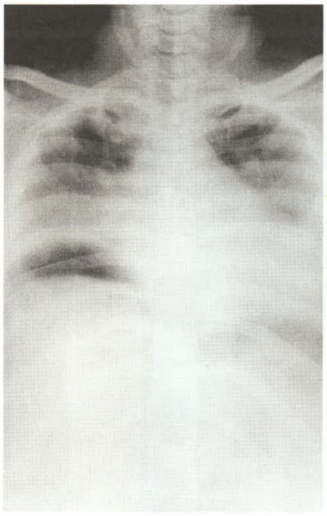


Fig. 27.24: Subphrenic abscess—mild pleural effusion on both sides

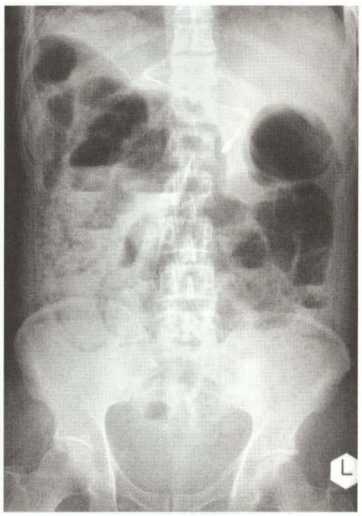


Fig. 27.25: Gas in the bowel wall indicates gangrene of the bowel

- Bile—green coloured—duodenum, stomach, gall bladder perforation
- Faeculent—dark green coloured thick aspirate with faecal odour—ileal perforations, postoperative anastomotic leaks
- Serous—exudative—early acute pancreatitis, tuberculous peritonitis
- Haemorrhagic—haemorrhagic pancreatitis
- Food particles—hollow viscus perforation

Thus ultrasound has so many advantages even though it may not point at the specific site. However, probe tenderness with fluid in the right iliac fossa may suggest acute appendicular perforation. Very thick contents such as anchovy sauce from ruptured amoebic liver abscess cannot be aspirated. However, ultrasound will give clue about the liver abscess.

5. Abdominal tap

- Aspiration of blood indicates haemoperitoneum or gangrene of the bowel.
- Aspiration of bile indicates biliary peritonitis due to perforation of duodenal ulcer, gall bladder or intestine.
- **Aspiration of frank pus** indicates peritonitis due to gram-negative bacteria. Foul-smelling pus is due to anaerobic bacteria producing free fatty acids and their esters. Always send the fluid for culture sensitivity (Fig. 27.26).
- Amylase estimation should be done to rule out pancreatitis.

6. Contrast-enhanced CT scan

- When the signs and symptoms are equivocal, CT is the ideal investigation.
- CT can diagnose hollow viscus perforation, especially when there is no gas under the diaphragm.
- CT can detect ischaemic changes due to gangrene of the bowel-gas in the bowel wall (Figs 27.27 and 27.28)

Fig. 27.26: Diagnostic tap showing pus following rupture of empyema of gall bladder

- CT can diagnose unsuspected and unexpected lesions in the abdomen including diverticular perforations, internal herniation and gangrene, acute pancreatitis, etc.
- Adequate hydration and normal renal function (as indicated by normal creatinine values) are important before a contrast-enhanced CT scan.

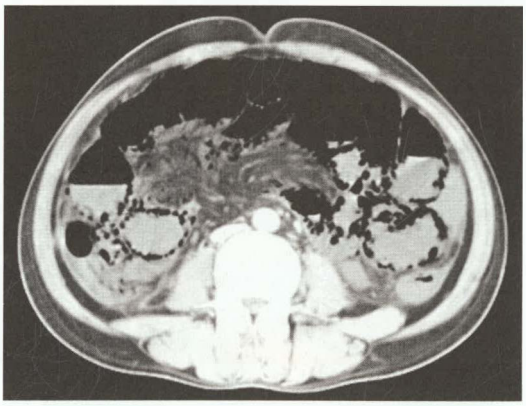


Fig. 27.27: CT showing air in the bowel wall

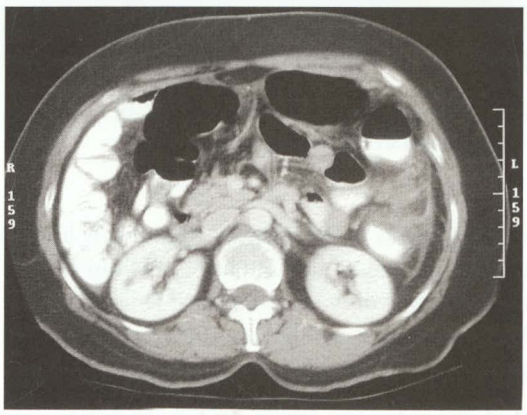


Fig. 27.28: CT showing hypodense lesion in the left iliac fossa—a case of sigmoid diverticular perforation

7. **Diagnostic laparoscopy** can be used in suspected cases of peritonitis (Key Box 27.6).

KEY BOX 27.6

DIAGNOSTIC LAPAROSCOPY

- It can be used to reconfirm peritonitis
- It can diagnose pancreatitis (laparotomy may be avoided)
- It can also treat primary cause, e.g. laparoscopic closure of duodenal ulcer perforation
- Peritoneal toilet can be given
- It can rule out other causes
- In blunt injury, **it can detect diaphragmatic injury**—herniation of bowel, etc.

PEARLS OF WISDOM

When in doubt, do laparoscopy: It can reveal 'hidden' pathology.

Treatment

1. **Aspiration:** Nasogastric aspiration with Ryle's tube helps in decreasing gastrointestinal secretion. Thus it reduces abdominal distension. It also prevents vomiting and gives rest to the gut. Indirectly it reduces 'bacterial load' contaminating peritoneum.
2. **Bowel care and blood:** Purgatives should not be given as it may result in perforation. Blood is arranged for surgery.
3. **Charts:** Temperature, pulse rate, respiratory rate, intake-output charts are maintained.
4. **Drugs** are given against gram-positive, gram-negative and anaerobic organisms (Key Boxes 27.7 and 27.8).

KEY BOX 27.7

SELECTION OF ANTIBIOTICS

- 2nd or 3rd generation cephalosporins should be started as early as possible
- Once culture and sensitivity reports are available (after surgery), antibiotics can be changed
- Antibiotics should also cover aerobes and anaerobes
- Should not have serious toxicity, especially amikacin which has nephrotoxicity. Hence, to be used carefully (check creatinine)

5. **Exploratory laparotomy** and appropriate surgery is done followed by thorough peritoneal toilet/wash with normal saline.
6. **Fluids**—IV fluids are given before, during and after surgery. Central venous cannulation and measurement of central venous pressure (CVP) is indicated in unstable patients to guide fluid therapy. If not possible, an ***emergency cut down*** (venesection)—cephalic or basilic vein, is done followed by fluid infusion. Preoperatively the aim is to maintain at least 30 ml/hr of urine output.
   - Ringer lactate solution is an ideal replacement.

KEY BOX 27.8

EARLY AGGRESSIVE RESUSCITATION

- **Restore** intravascular volume. **Crystalloids:** Ringer lactate or isotonic saline stay in the intravascular space for a short period, larger volumes required
  Colloids: Longer duration of action, smaller volumes are sufficient and can be used in cardiac patients
- **Restore** oxygenation by face mask or mechanical ventilation, as necessary
- **Restore** perfusion: Dopamine/dobutamine/noradrenaline.
- **Restore** normality by 'war' against sepsis—**ANTIBIOTICS** and surgical removal of **SEPSIS**

Principles of surgery for peritonitis

(Key Box 27.9, Figs 27.29 to 27.33)

1. Generous **incision** is used
2. As soon as the peritoneal cavity is opened, purulent fluid comes out. The fluid is collected and sent for **culture** and **sensitivity**. Greenish fluid indicates a hollow viscus perforation. All the fluid is drained, the source of peritonitis is identified and appropriate surgical procedure is done.
   Examples are
   - Appendicectomy for appendicitis.
   - Closure of perforation for perforated peptic ulcer.
   - Closure or resection for ileal perforation.
   - Resection of the bowel for gangrene.

   Control of sepsis: This is the most important step of treatment of peritonitis. Removal of septic focus is a primary aim—examples: Appendicectomy, perforation closures (dudodenal ulcer) or resection (intestinal or colonic perforation) or cholecystostomy in difficult perforated gall bladder diseases. However, all the septic foci in the abdomen have to be removed—necrotic material, pus pockets and food particles. Thorough irrigation with warm saline cleans up subhepatic spaces, pelvic spaces and interloop collections. Primary anastomosis in presence of sepsis may

KEY BOX 27.9

PRINCIPLES OF SURGERY

- Incision
- Establish the diagnosis
- Exploration
- Pus culture and sensitivity
- Treat the cause—control of sepsis
- Peritoneal toilet
- Drain
- Closure

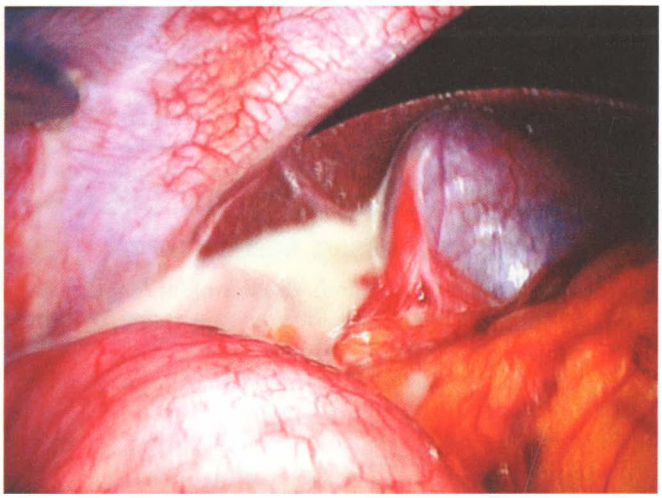


Fig. 27.29: Laparoscopy showing pus

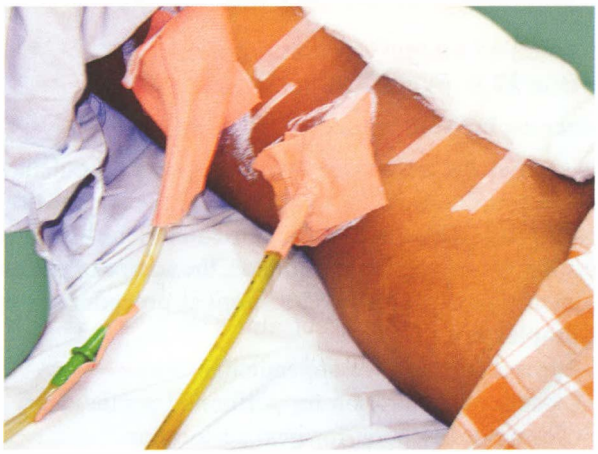


Fig. 27.30: A late case of duodenal ulcer perforation who presented with peritonitis with sympathetic pleural effusion. Laparotomy, closure of perforation and feeding jejunostomy were done. Drains in the subhepatic space and right pleural cavity were placed. Patient had stormy postoperative period with biliary fistula, which was closed after 6 weeks

result in leak and postoperative peritonitis. It is better to do colostomy or ileostomy in such cases. Incision can be partially closed leaving the skin open—sutures can be tied after 2 days in the ward. In a few cases, laparostomy is done if you suspect abdominal compartment syndrome.

3. It is better to use **nonabsorbable suture** material such as silk to do an intestinal anastomosis or for closure of perforation. In the presence of infection, absorbable sutures such as catgut get absorbed very fast.
4. A thorough **peritoneal wash/lavage** is given by using warm saline (up to 3–5 litres) to avoid intraperitoneal abscesses. Antiseptic agent such as betadine solution should be avoided because they can cause adhesions (Key Box 27.10).
5. Peritoneal cavity is drained to the exterior by using **tube drains**. These are kept in the subhepatic space and in the pelvic cavity.

KEY BOX 27.10

PERITONEAL LAVAGE

- Used in diffuse peritonitis
- 3–5 litres of isotonic crystalloid solution is used.
- Avoid antibiotic solution or povidone iodine solutions—they may induce more adhesions.
- Aminoglycoside lavage may cause respiratory depression due to neuromuscular blocking action of these drugs. Mops must be used to dry the peritoneal cavity. If fluid is left over, it may dilute the opsonins and thus decrease phagocytosis.

Fig. 27.31: Tension sutures

Fig. 27.32: Zip closure of peritoneum

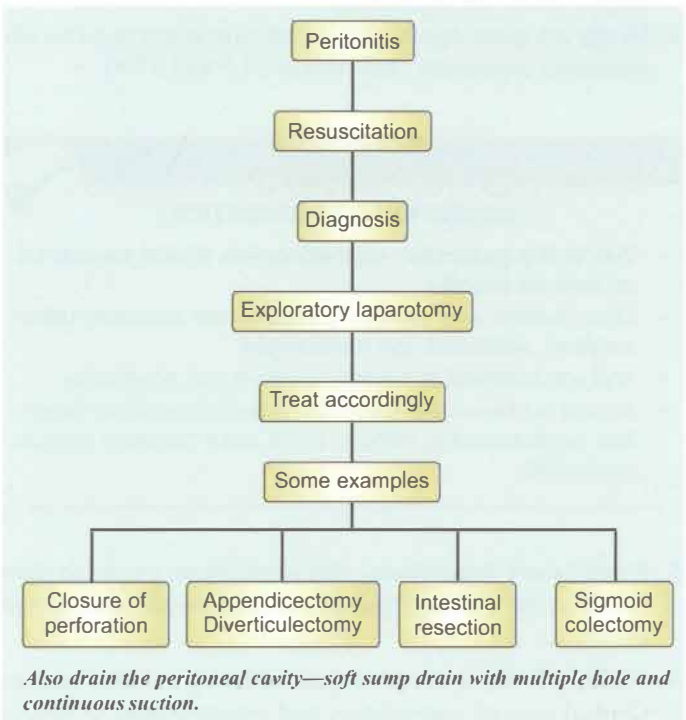


Fig. 27.33: Principles of management of peritonitis—identifying the source of sepsis and elimination is the key to success

6. The wound is irrigated with **antiseptic agents**.
7. **Tension sutures** are put depending upon the severity of the peritonitis to prevent burst abdomen.
8. **Laparostomy** ***(vide infra)*****:** This method of exposing the peritoneal cavity can be done in selected cases, if you suspect abdominal compartment syndrome.

LAPAROSTOMY

This refers to leaving peritoneal cavity exposed to outside without approximation of the anterior abdominal wall. Some situations arise, especially in emergency cases, where this is required. Hence, it is important to know how to deal with this situation.

Types

1. **Open laparostomy:** Abdominal fascia and peritoneum are not sutured.
   Advantages: Abdominal compartment syndrome can be prevented. Details are given later.
   Disadvantages: Significant fluid loss and secondary infection.
2. **Closed laparostomy or mesh laparostomy:** Here the fascial layer is closed by using marlex mesh or prolene mesh or even a zip to protect exposed viscera.
   Advantages: One can minimise infection.
   Disadvantages: Abdominal compartment syndrome and perforation of bowel can occur.

Indications of laparostomy

When a second look procedure is contemplated, e.g. acute pancreatitis, mesenteric ischaemia.

Nonoperative treatment

1. Too sick a patient to tolerate the surgical procedure.
2. Sealed perforation
3. Localised peritonitis—may resolve with treatment.

ABDOMINAL COMPARTMENT SYNDROME

Introduction

The phrase "abdominal compartment syndrome" was coined in 1984 when Irving Kron, described the measurement of intra-abdominal pressure as a means of developing criteria for abdominal decompression to improve organ function.

Definition

- Abdominal compartment syndrome (ACS) is defined as a sustained increase in IAP more than 20 mmHg [with or without an abdominal perfusion pressure (APP) < 60 mmHg)] that is associated with new organ dysfunction/ failure (Fig. 27.34).

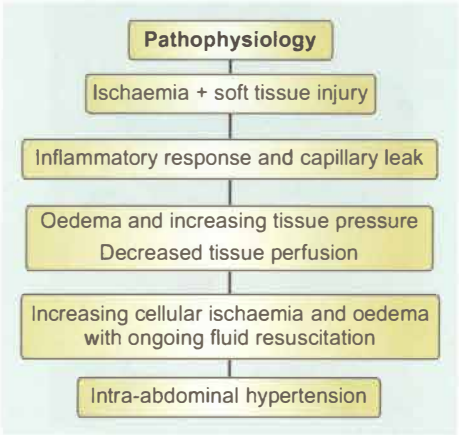


Fig. 27.34: Pathophysiology of abdominal compartment syndrome

- Intra-abdominal pressure (IAP) is the pressure concealed within the abdominal cavity. Normal IAP is 0–5 mmHg showing phasic variation with respiration.
- Intra-abdominal pressure is measured to detect abdominal compartment syndrome and to decide on the requirment of a decompression so as to improve organ function.

Final effects

- The adverse physiological effects of intra-abdominal hypertension (IAH) affect almost every organ system resulting in ACS. The major systems affected, in decreasing frequency of incidence and morbidity are: Pulmonary, cardiovascular, renal, splanchnic, central nervous system. Thus the end result can be:
  - Intractable hypoxia, hypercarbia, ARDS
  - Cardiac insufficiency and cardiac arrest
  - Oliguria, anuria, acute renal failure
  - Cerebral oedema and anoxia

Grading of IAP (Burch)

Grade I : IAP 12–15 mmHg
Grade II : IAP 16–20 mmHg
Grade III : IAP 21–25 mmHg
Grade IV : IAP > 25 mmHg

Risk factors

A. Diminished abdominal wall compliance

1. Acute respiratory failure with elevated intrathoracic pressure.
2. Abdominal surgery with primary fascial or tight closure. Example: Reduction of massive hernia (Figs 27.35 and 27.36).
3. Major trauma/burns
4. Prone positioning, head of bed >30°
5. High BMI, central obesity

Fig. 27.35: This boy had abdominal compartment syndrome after reduction of intestinal contents from left thoracic cavity and we could not close the abdomen. It was covered with a thin plastic sheet—you can see the intestine. It took two months for granulation tissue to cover the defect. Eventually, he recovered with an incisional hernia. Bogota bag or VAC are the other alternative methods of closure

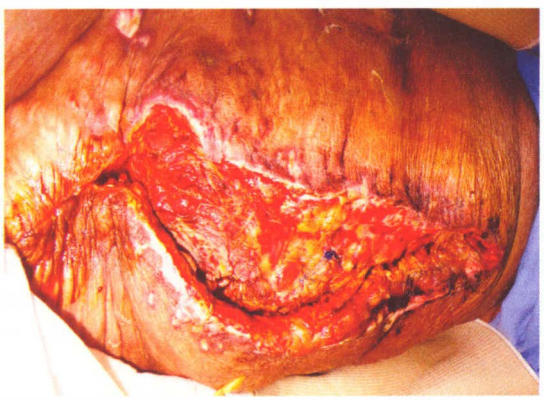


Fig. 27.36: Loss of abdominal layers following reduction of a massive ventral hernia

B. Increased intraluminal contents

1. Gastroparesis
2. Acute gastric dilatation
3. Ileus
4. Colonic pseudo-obstruction

C. Increased abdominal contents

1. Haemoperitoneum/pneumoperitoneum
2. Ascites/liver dysfunction
3. Laparoscopy

IAP measurement—principles

- Expressed in mm Hg (1 mm = 1.36 cm of water).
- Measured at end expiration
- Performed in supine position
- Zeroed at mid-axillary line at the level of iliac crest.
- Performed with an instillation volume of no greater than 25 ml of saline (for bladder technique).
- Measured 30–60 secs after to allow bladder detrusor muscle relaxation (for bladder technique).

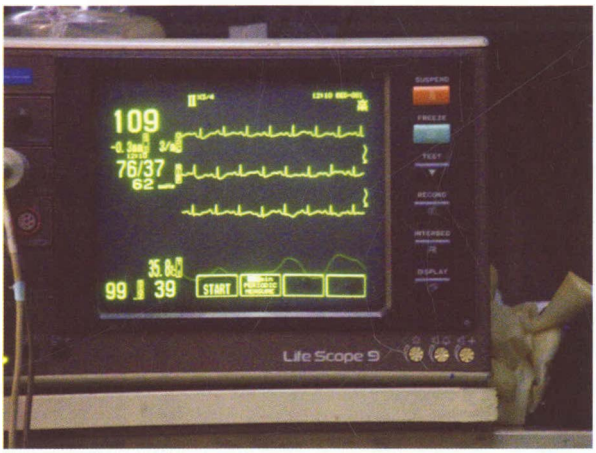


Fig. 27.37: Close monitoring of patient with vital signs and blood transfusion in addition to antibiotics play a major role in the management of septic shock

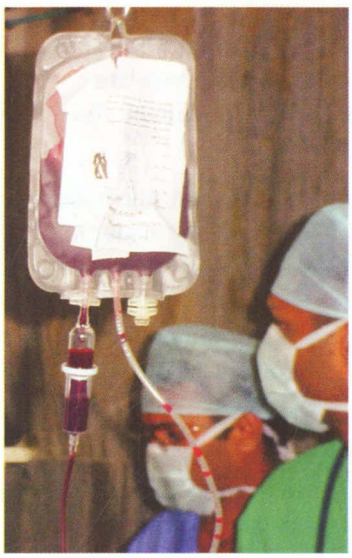


Fig. 27.38: Blood transfusion

TREATMENT

- Close monitoring of patient's vitals (Fig. 27.37)
- Blood transfusion when required (Fig. 27.38)

SURGERY

I. Temporary abdominal closure

- Towel clip closure, only skin closure.
- Mesh—commercially available meshes with absorbable surface facing intraperitoneum and non-absorbable facing outer aspect of the wound are used. Example being polygalactin (vicryl inside) and polypropylene (prolene) outside can be used.
- **PTFE mesh repair:** Expanded polytetrafluoroethylene. (ePTFE) is another mesh which is used. It is a surgical biomaterial with two antimicrobial preservative agents—chlorhexidine diacetate and silver carbonate. It also enhances tissue ingrowth.

CLINICAL NOTES

A young boy of 18 years, who suddenly had breathlessness was found to have diaphragmatic hernia. After reduction and closure, patient developed ACS. Reopening of abdomen was done and peritoneum was not closed but a 'cover' was given by using 'urosac' bag which was split open. Wound was allowed to heal by granulation tissue.

- **Bogota bag:** A **Bogota bag** is a sterile plastic bag used for closure of abdominal wounds.[1] It is generally a sterilised, 3 litre genitourinary irrigation bag that is sutured to the skin or fascia of the anterior abdominal wall. The Bogota bag acts as a hermetic barrier that avoids evisceration and loss of fluids. Another advantage to the Bogota bag, is that the abdominal contents can be visually inspected which is particularly useful in cases of ischaemic bowel. Thus useful in resections following mesenteric ischaemia. In our country we can use urosac bag (which can be split open) or even a thin plastic sheet can be used like a Bogota bag (Fig. 27.35).
- **Vacuum assisted closure (VAC):** It has been extensively used in the management of leg ulcers specially diabetic ulcers. It has been used in a few cases of severe pancreatitis. Here it is called open abdomen negative pressure therapy system. It removes debris, inflammatory exudates.

II. Definitive abdominal closure

- Primary closure. It is done layer by layer. Non-absorbable suture is usually selected
- Synthetic mesh
- Biologic mesh
- Component separation
- Plastic surgery

PEARLS OF WISDOM

No doubt, drainage of the septic focus is the most important step.

COMPLICATIONS OF PERITONITIS

1. **Severe hypovolaemic shock** giving rise to renal failure. It can be prevented by adequate hydration of the patients and careful usage of antibiotics such as gentamicin.
2. **Septic shock, multiorgan failure** and death occur in late cases of peritonitis.
3. **Subacute intestinal obstruction** due to postoperative adhesions.
4. **Pelvic abscess**
5. **Subphrenic abscess**

[1]Bogota bag's use was first described by Oswaldo Borraez in Bogota, Colombia.

PELVIC ABSCESS

This refers to accumulation of pus in the rectovesical pouch or pouch of Douglas (rectouterine pouch).

Causes

- **Any peritonitis,** commonly following perforation due to ***acute appendicitis*** or following salpingo-oophoritis. The rectovesical pouch is the most dependent part in the body. Hence, the septic emboli accumulated in peritoneal space give rise to pelvic abscess.
- **Anastomotic leakage** is also an important cause.
- Perforated duodenal ulcer, perforated ileal ulcers on the other common causes.

Clinical features

- History of surgery/peritonitis
- Postoperative ***high-grade fever***
- History of **discharge of mucus per rectum** for the first time in a patient who is **recovering from peritonitis** suggests pelvic abscess. It occurs due to irritation of the rectum. Increased frequency of micturition occurs due to irritation of bladder.
- Deep tenderness in the suprapubic region.
- Surgery done for peritonitis
- Continuing infection even after surgery—leak from an anastomotic line.
- Inadequate peritoneal toilet
- Inappropriate antibiotics
- Pelvic surgery
- Diabetes

CLINICAL NOTES

A 36-year-old lady underwent vaginal hysterectomy for dysfunctional uterine bleeding. To control the bleeders, several gauze pieces were used without a proper count. After 2 weeks, purulent discharge per vagina, fever, ill health was reported. Ultrasound done showed a pelvic abscess. CT scan done in our hospital showed foreign body with air trap suggesting gauze pieces. Her abdomen was explored, abscess drained and the gauze pieces were removed.

Diagnosis

- Confirmed by **per-rectal examination**. A **tender boggy** swelling is felt in the anterior wall of rectum. **Ultrasound** can define an abscess and can detect the size of the abscess.
- **CT scan** is very useful in defining pelvic abscess, its extent and to detect presence of a foreign body (Fig. 27.39).

Fig. 27.39: CT scan showing gauze pieces—she had persistent foul-smelling vaginal discharge

Treatment (Fig. 27.40)

- Under general anaesthesia, a proctoscope is introduced and a nick is made in the anterior wall of the rectum to open into the abscess cavity. Pus is drained with a sinus forceps through the rectum. There is no peritoneal contamination. The cavity collapses after a few days. Postoperatively, the patient is given broad spectrum antibiotics.
- In females, pus can be drained through posterior fornix.

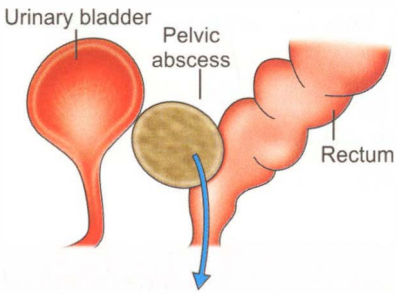


Fig. 27.40: Drainage of pelvic abscess through the rectum

SUBPHRENIC ABSCESS

Introduction

- As a result of peritonitis, residual abscess can collect in the intraperitoneal cavity. Pus that collects under the diaphragm is described as subphrenic abscess. Subphrenic abscess is the commonest intra-abdominal abscess.
- Gastrointestinal perforations, postoperative leaks, penetrations, traumas, puerperal sepsis, are the common causes of subphrenic abscess.
- Blood clots, bacteria laden fibrin, neutrophil contribute to an abscess.

Surgical anatomy

There are 5 subphrenic spaces between the diaphragm and the liver bounded by various peritoneal folds. Four are intraperitoneal and one is extraperitoneal. The spaces, boundaries and the common causes of pus in these spaces are described in Table 27.1 and Figs 27.41 to 27.43.

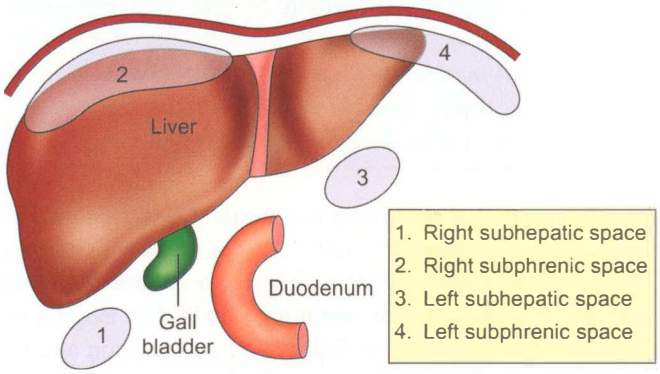


Fig. 27.41: Subphrenic spaces in sagittal section

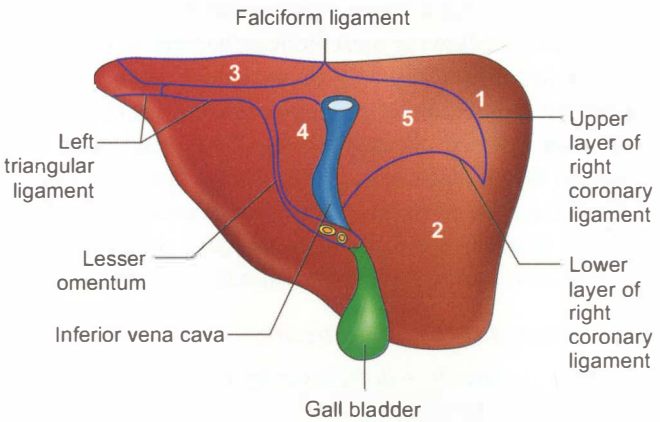


Fig. 27.42: Subphrenic spaces (Table 27.1 for numbers)

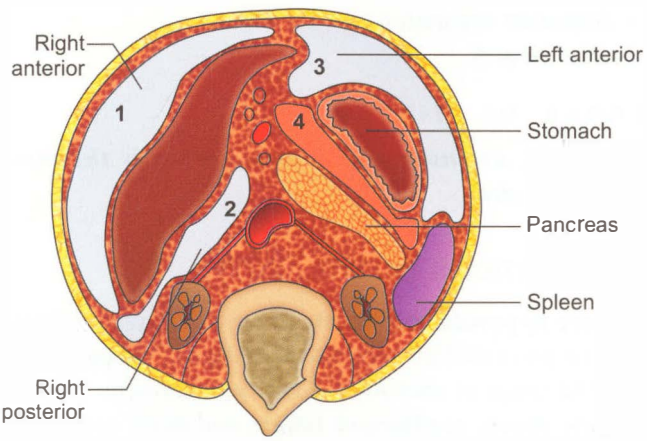


Fig. 27.43: Cross-section showing subphrenic spaces: (1) Right anterior intraperitoneal space, (2) right posterior intraperitoneal space, (3) left anterior intraperitoneal space, (4) left posterior intraperitoneal space (lesser sac)

Aetiopathogenesis (Fig. 27.44)

- The causative organisms of peritonitis are *Escherichia coli*, Enterococci, Klebsiella, Enterobacter, Proteus, Bacteroides, etc.

Table 27.1 Subphrenic space boundaries and common causes of involvement

Spaces	Boundaries	Common causes
1. Right anterior intraperitoneal space	It lies between right lobe of the liver and diaphragm Posteriorly—anterior layer of the coronary ligament and right triangular ligament. On the left side is the falciform ligament	Perforated duodenal ulcer, gastric ulcer, cholecystitis
2. Right posterior intraperitoneal space (Rutherford Morrison's hepatorenal pouch)	It lies below the right lobe of the liver Inferiorly—hepatic flexure and transverse colon Medially—second part of the duodenum Laterally—abdominal wall. This is the biggest intraperitoneal space	Appendicitis, cholecystitis Perforated duodenal ulcer Upper abdominal surgery
3. Left anterior intraperitoneal space	Above—diaphragm. Posteriorly—left triangular ligament, left lobe of the liver, lesser omentum, anterior surface of the stomach. Right side—falciform ligament Left side—spleen	Surgery on stomach—gastrectomy. Distal pancreatectomy. Left hemicolectomy
4. Left posterior intraperitoneal space (lesser sac)	In front—by lesser omentum and posterior surface of the stomach. Behind—pancreas, suprarenal, left kidney On the right side—foramen of Winslow through which it communicates with the greater sac	Pseudopancreatic cyst Perforated gastric ulcer
5. Midline extraperitoneal space (bare area of the liver)	Above—upper layer of coronary ligament Below—lower layer of coronary ligament Left—inferior vena cava	Amoebic hepatitis Pyogenic liver abscess

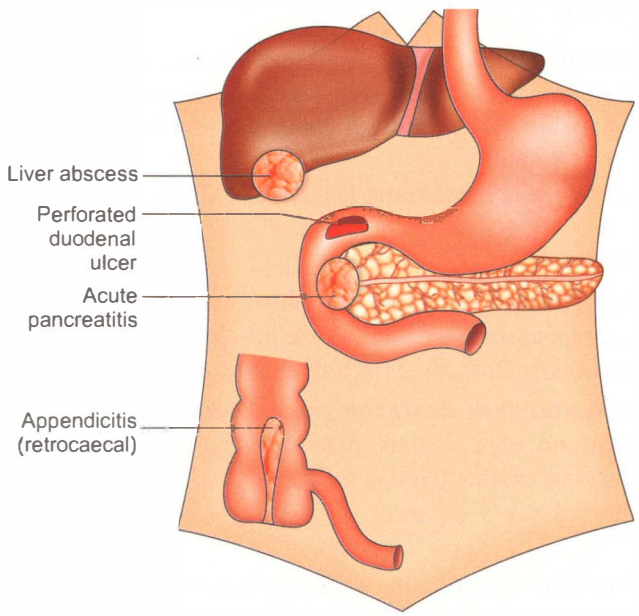


Fig. 27.44: Common causes of subphrenic abscess

- The high incidence of subdiaphragmatic abscess is due to constant circulation of fluid from below upwards because of following reasons:
  1. Upward movement of diaphragm during expiration.
  2. Decreased intra-abdominal pressure
  3. Capillary action

Subphrenic abscess is common on the right side because of the following reasons:

1. Right paracolic gutter is wide and deep and colophrenic ligament is absent.
2. Left paracolic gutter is narrow and colophrenic ligament is present on the left side.
3. Majority of diseases affect right side (perforation, liver abscess, appendicitis, gall bladder disorders, etc.) (Fig. 27.44).

Clinical features

- A patient who is recovering from peritonitis complains of ***fever with sweating***. Initially, fever is low grade, continuous. Later, there is high-grade fever with chills and rigors.
- **Deterioration of health** occurs very fast with wasting and anorexia.
- **Shoulder pain** is due to irritation of undersurface of the diaphragm by the pus (sensory fibres of the phrenic nerve are irritated—C3, 4).
- Postoperative patient is not doing well—prolonged ileus.
- Anorexia, wasting, hiccup, dry cough.

PEARLS OF WISDOM

A postoperative patient who has pyrexia, prolonged ileus, poor appetite and progressive deterioration of health has subphrenic abscess.

- **Tenderness** is present in the epigastrium on deep palpation.
- Common causes of postoperative fever are absent, e.g. thrombophlebitis, urinary tract infection.

PEARLS OF WISDOM

Pus nowhere, pus somewhere, pus under the diaphragm—Harold Barnard.

Investigations

1. **Total count** with neutrophil count
2. **Plain X-ray abdomen** (erect)—may show gas and fluid level under the diaphragm (Figs 27.45 and 27.46)
3. **Fluorescent radiography** may reveal absence of movement of right side of diaphragm on inspiration.
4. **Ultrasonography** confirms the site of abscess, number of abscesses, loculations, etc.
   - Abscess is characterised by hypoechogenic cavity surrounded by sharp distinct echogenic wall. It can be therapeutic to insert catheter for drainage.

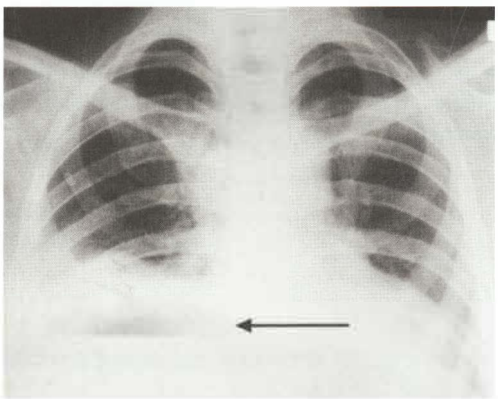


Fig. 27.45: Plain X-ray—gas and fluid level under diaphragm: PA view and lateral view

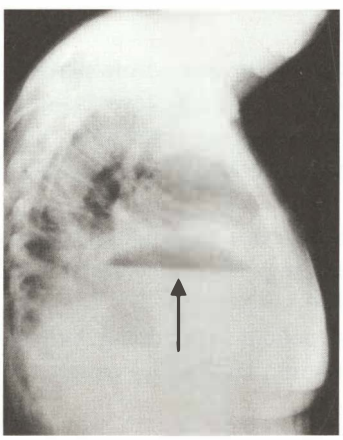


Fig. 27.46: Plain X-ray—gas and fluid level under diaphragm: PA view and lateral view

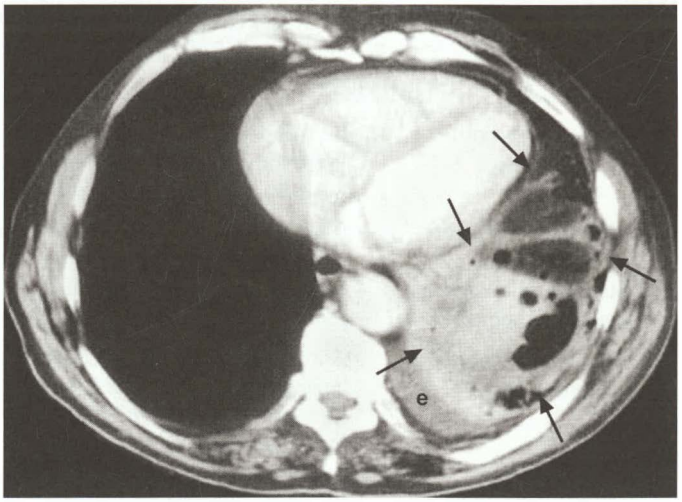


Fig. 27.47: CT scan showing left subphrenic collection following acute pancreatitis—a pigtail catheter is inserted (also *see* Fig. 27.48)

5. **CT scan** demonstrates well defined, low density mass, the rim of which is enhanced after intravenous injection of contrast medium. The mass tends to be round because of centripetal expansion—highly sensitivity > 95% (Fig. 27.47).
6. **Isotope imaging** using Gallium 67 citrate or Iridium 111. Gallium binds to proteins—**lactoferrin and transferrin** which are present in **high concentration** in an abscess.

Goals of the treatment (Flowchart 27.2)

Treatment

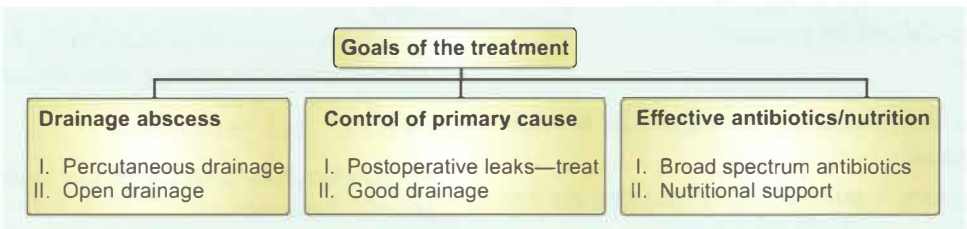
Today with the availability of sophisticated imaging facilities, percutaneous drainage has become the choice of therapy rather than surgery. Both have been described in next column.

I. Percutaneous drainage can be done with the help of ultrasound or CT scan, provided the abscess cavity is unilocular, and the track is safe.

Types

1. **Pigtail catheter** (using Seldinger's technique): It is a small tube used to drain bile, urine, pancreatic fluid or abscess.
2. **Trocar catheter:** 12–16 F trocar is used.

Flowchart 27.2



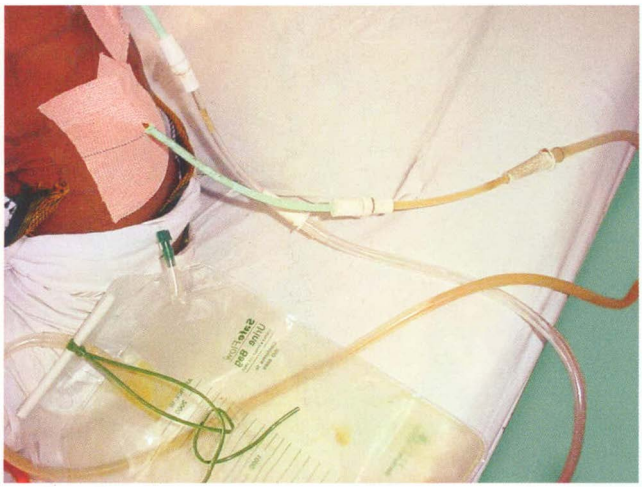


Fig. 27.48: Pigtail catheter drainage of left subphrenic abscess following gastric ulcer perforation

3. **Sump catheter:** It has a double lumen which permits irrigation as well as drainage and allows a good suction (Key Box 27.11).

PEARLS OF WISDOM

More than 90% of subphrenic abscesses are managed by percutaneous drainage successfully.

KEY BOX 27.11

INDICATIONS FOR REMOVAL OF CATHETER

- Drainage less than 10 ml/day
- No fever, no pain
- WBC counts return to normal

II. Open drainage (Key Box 27.12)

- The anterior subcostal or posterior (bed of 12th rib) approach is used. Both are extraserous approaches.
- However, lesser sac abscess and abscesses connected with the bowel, discharging pus or bile, are drained by intraperitoneal route.
- Open drainage is ideal in cases of multiloculated abscesses.
- Surgery is always done under the cover of broad-spectrum antibiotics.

KEY BOX 27.12

INDICATIONS FOR OPEN DRAINAGE

1. Multiloculated abscess
2. Persistent fistula discharging pus—communication with bowel.
3. Thick viscid content
4. Failure of percutaneous aspirations
5. Abscess very close to IVC/diaphragm

SPECIAL TYPES OF PERITONITIS

POSTOPERATIVE PERITONITIS

Introduction: This is not an uncommon problem encountered in the surgical wards. Very often patient who undergoes an intestinal or a biliary surgery and a few days later vague symptoms and signs develop. Hence it is difficult to diagnose if one takes a casual approach. It has high mortality. Hence requires early detection and demands early effective solution.

It should be suspected following surgery on intestines or biliary tract, when a patient who is recovering from paralytic ileus starts deteriorating or when paralytic ileus does not return back to normal.

Etiology (Figs 27.49 to 27.52)

- Leakage from anastomotic line
- Iatrogenic visceral trauma
- Foreign bodies
- Others

Causes of delay in the diagnosis

- Presence of fever is attributed to other sources of infection such as urinary tract infection, thrombophlebitis, etc.
- Presence of pain and tenderness is attributed to recent laparotomy scar.
- Tachypnoea, hypotension are attributed to pre-existing medical conditions such as COPD, cardiac failure.
- Steroid therapy masks the local signs and symptoms.
- Administration of antibiotics would have reduced the severity of peritonitis (masking effect) only to manifest as septicaemia some time later.

Bacteriology

1. Common organisms in postoperative peritonitis (Figs 27.49 to 27.52)

A. Gram –ve Bacilli	*E. coli*
Klebsiella	Proteus
Pseudomonas	
B. Anaerobes	Bacteroides
Clostridium	Peptostreptococci
Fusobacteria	
C. Gram +ve cocci	Enterococci
Staphylococci	Streptococci

2. Foreign bodies responsible for postoperative peritonitis

Macroscopic	Microscopic
• Gossypiboma	• Barium
• Textiloma	• Cloth piece
• Surgical drains	• Faecal matter
• Suture materials	• Necrotic tissue
• Surgical clips	• Talcum powder
• Implants	
• Instruments	

3. Presentation of foreign body

• Abdominal pain	• Intestinal obstruction
• Mass abdomen	• Fistula and sinus
• Granuloma, fever	• Extrusion—sometimes the foreign body may be visible to the exterior

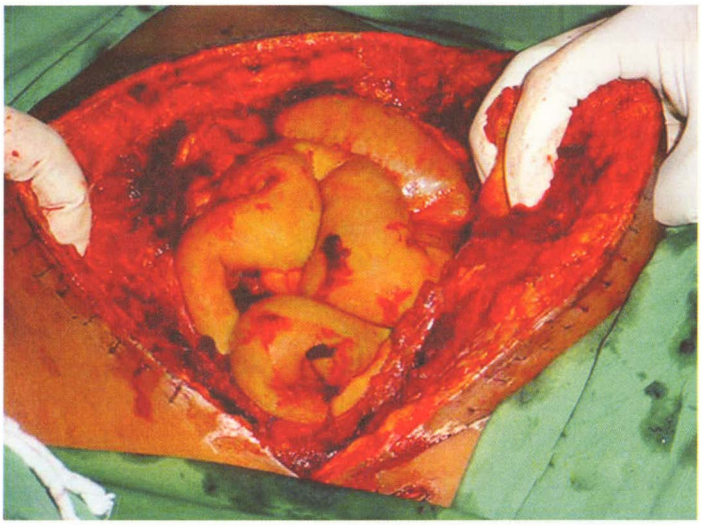


Fig. 27.49: Postoperative peritonitis due to post-cholecystectomy leak

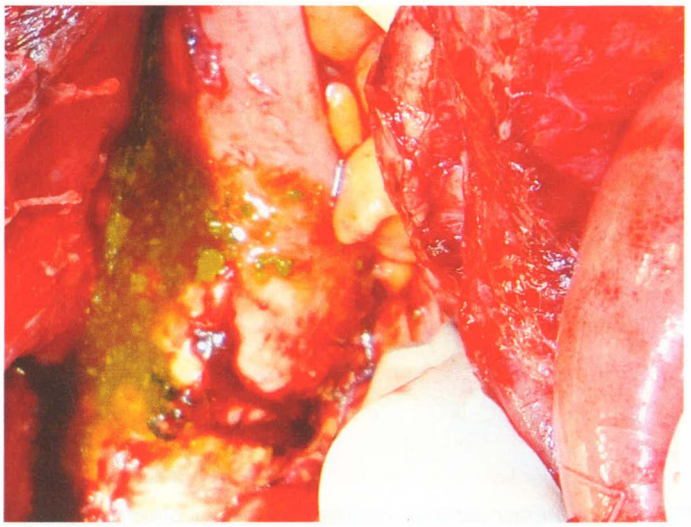


Fig. 27.50: Postoperative peritonitis due to anastomotic leak with faecal peritonitis

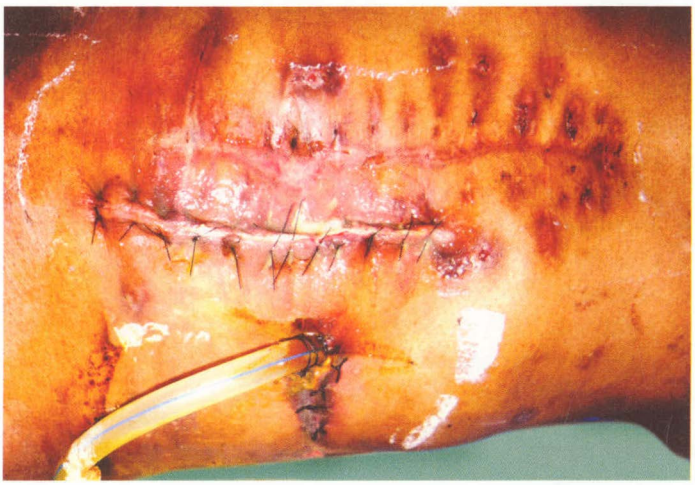


Fig. 27.51: Postoperative peritonitis due to enteric ulcer perforation

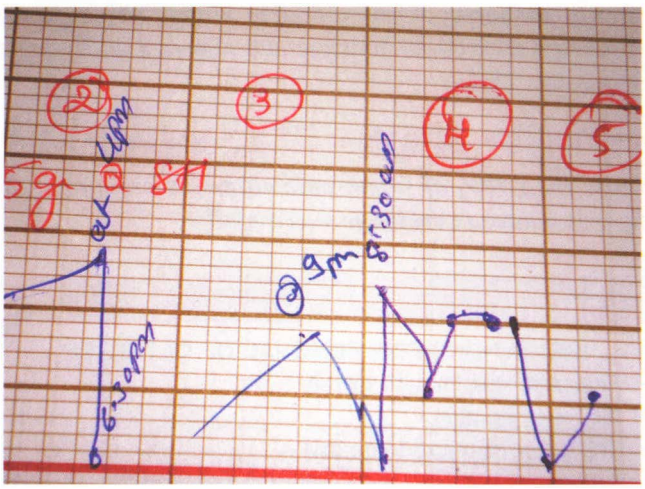


Fig. 27.52: Postoperative peritonitis due to anastomotic leak with faecal peritonitis—look at the temperature chart

How to suspect postoperative peritonitis

- Deterioration after 3–5 days of operation (the time when anastomotic dehiscence takes place).
- Delay in recovery from paralytic ileus—abdominal distension.
- Evidence of toxaemia—tachycardia, tachypnoea.
- Free drainage of bile and faecal matter or pus from the drain site or the main wound.
- Oliguria may be an early indicator of postoperative sepsis.
- Tenderness, guarding, rigidity.

Treatment (Figs 27.53 and 27.54)

1. Prevention of gossypiboma (MOP)
   - Double counting
   - Sponges with radio-opaque markers
   - No hurried counting
   - Additional counting—when change of OT personnel
   - Avoid using packs—fascial closure
   - Intraoperative radiology
   - High degree of suspicion (*see* clinical notes)
2. Specific treatment (Key Box 27.13)

CLINICAL NOTES

In 1973, when ultrasound facilities were not available, a 35-year-old male who had undergone appendicectomy for a gangrenous appendix, was found to have high spiking fever on the 3rd postoperative day. All possible causes of postoperative fever including malaria were ruled out. On the 10th postoperative day, the patient developed a purulent discharge of 100 ml through the lower part of the main wound following which he had a spontaneous and dramatic recovery.

KEY BOX 27.13

TREATMENT

- Danger lies in delay, not in reoperation
- Leak or abscess cavity is confirmed by abdominal ultrasound.
- Exploration (preparation similar to a routine laparotomy)
- Resection or resuturing, ileostomy, colostomy
- Drainage of abscess cavity
- Refashioning of colostomy, if it is retracted
- Once abscess is drained or leakage is prevented, recovery is wonderful.
- Appropriate antibiotics
- Delayed closure of skin
- Peritoneal lavage

Fig. 27.53: Pus pouring out as soon as peritoneum is opened. First step is to send a pus for culture sensitivity

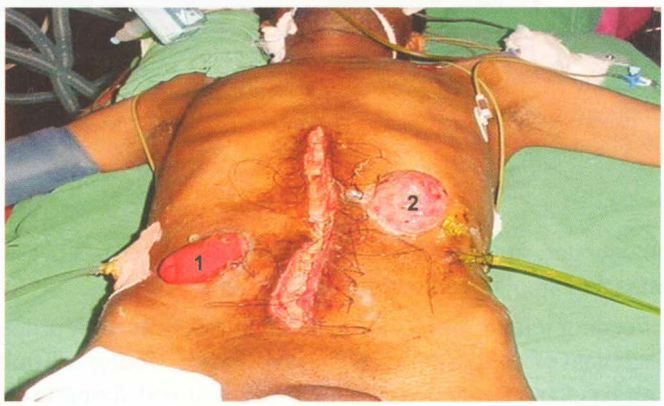


Fig. 27.54: Postoperative peritonitis following due to leak following right hemicolectomy for carcinoma caecum. There were 2 sites of leak: 1. Ileocolic anastomotic site, 2. proximal jejunum. Both were exteriorized. You can see the wound left open with loose skin sutures—they will be tied after 3–4 days. This is to prevent wound infection (almost inevitable)

Poor prognostic factors in postoperative peritonitis

- Increasing age
- Organ(s) failure
- Colonic perforation
- Multiple abscess
- Lesser sac abscess
- Malnutrition
- Postoperative pneumonia
- **Anergy:** Anergy (immunologic tolerance) refers to the failure to mount a full immune response against a target.

BILIARY PERITONITIS

- Leakage of bile into the peritoneal cavity results in biliary peritonitis.
- It will be more obvious and can be detected early if a drainage tube has been kept.

Causes

1. Surgery on the gall bladder
- Leakage from the cystic duct
- Injury to the right hepatic duct
- Leak from accessory cholecystohepatic duct

2. Surgery on the CBD
- Retained stones in the lower CBD
- Loose sutures over CBD
- T-tube not anchored properly

3. Surgery on the duodenum
- Sphincteroplasty
- Partial gastrectomy
- Reperforation of sutured duodenal ulcer

4. Injury to the duodenum
- During nephrectomy, hemicolectomy
- Blunt injury

5. Instrumentation

ERCP, stenting or following duodenal polypectomy

6. Diseases of the gall bladder

Perforation or gangrene of the gall bladder

Clinical features (Figs 27.55 and 27.56)

- In majority of the cases, the local signs are confined to one quadrant of the abdomen in the form of guarding and rigidity.
- There may be excoriation of the skin due to drainage of bile to the exterior (Fig. 27.57)
- However, when the anastomosis gives way, generalised peritonitis can occur.
- In untreated cases, septicaemic shock can develop.
- Final stage will be multiorgan failure and death.
- It has a very high mortality rate.

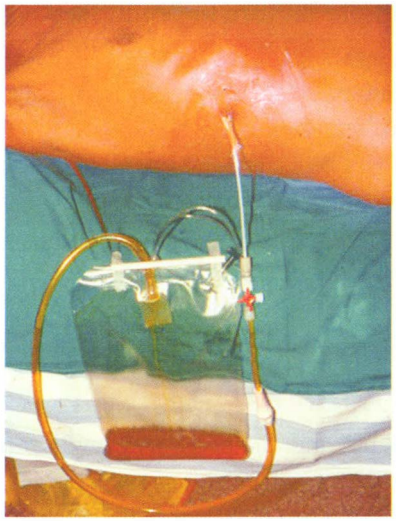


Fig. 27.55: Post-cholecystectomy bile leak due to injury to the common bile duct. Majority of such leaks can be managed conservatively. If the leak continues, ERCP and treat the cause accordingly

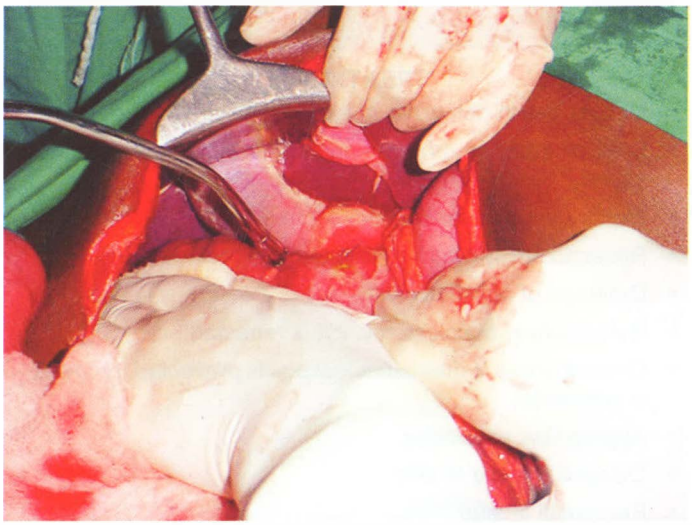


Fig. 27.56: Biliary peritonitis following duodenal ulcer perforation. It requires urgent laparotomy. Closure of the ulcer perforation, peritoneal lavage followed by drainage tube and closure of the incision

Fig. 27.57: Injury to the terminal ileum during tubectomy resulted in faecal fistula. To prevent the excoriation of the skin, zinc oxide cream has been applied

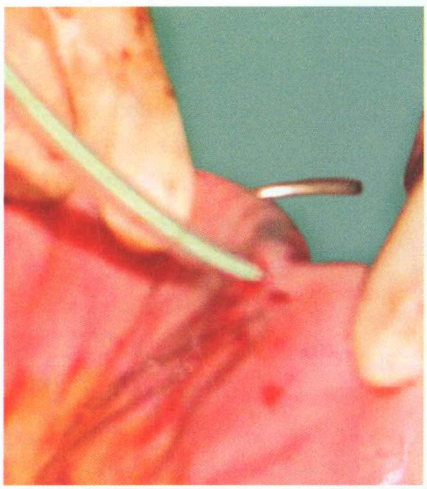


Fig. 27.58A: Feeding jejunostomy—a small incision is made in the proximal jejunum and a plastic tube is introduced into the lumen and advanced to about 60 cm

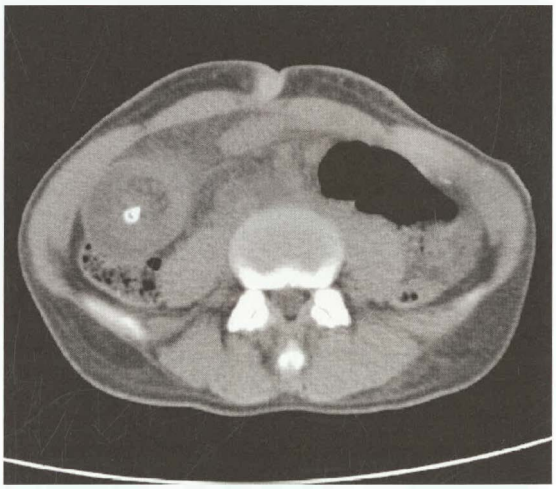


Fig. 27.58B: Feeding jejunostomy intussusception

Treatment

1. Most of the biliary fistulae heal within 2–3 weeks with conservative line of treatment.
2. If it does not heal, re-exploration and resuturing or resection has to be done.
3. Feeding jejunostomy is a very useful procedure in all cases of reperforation of sutured duodenal ulcers or difficult duodenal ulcer closures (Fig. 27.58).

It should be remembered that feeding jejunostomy is temporary and is get till it is assured that there is no leak from the operated site. If there is no peritonitis, patient has passed flatus and stools, jejunostomy tube is removed.

Feeding jejunostomy should be done carefully with proper placement of catheter within the small bowel, fixing it firmly both inside and outside the abdominal wall and fixing to the bowel wall.

Complications of feeding jejunostomy

- Bile leak from the entry point
- Tube blockage if it is not flushed properly after feeding.
- Bleeding, displacement and intussusception.

PNEUMOCOCCAL PERITONITIS

- Primary variety is more common. Girls of 3–6 years of age are usually affected. Infection spreads from female genital tract through vagina.
- Malnourishment precipitates pneumococcal peritonitis.
- In boys, blood spread can occur following upper respiratory tract infection.

Clinical features

- High grade fever with features of toxaemia.
- Bloody diarrhoea and frequency of micturition are indicative of pelvic peritoneal inflammation.
- Other features of peritonitis are present.

Diagnosis

Aspiration of peritoneal fluid demonstrates high WBC count—30,000/mm^3. More than 90% are polymorphs.

Treatment

Laparotomy and drainage of pus (odourless and sticky initially and creamy or purulent in the later stages), to be followed by appropriate antibiotics.

PRIMARY STREPTOCOCCAL PERITONITIS

- Infants and children less than 4 years are commonly affected.
- Peritoneal exudate is cloudy and contains flakes of fibrin.
- Symptoms of gastroenteritis—greenish watery stools are present.
- Source of infection is tonsillitis, pharyngitis, etc.
- Treated with injection crystalline penicillin.

PARTURITION PERITONITIS

- This occurs after delivery if proper aseptic precautions are not taken. The incidence has come down in the recent years. Attempted abortions by using instruments which are not sterile results in peritonitis. Most of the time, peritonitis is confined to pelvis with paralytic ileus, mucous diarrhoea and offensive lochia (Key Box 27.14). Late cases develop generalised peritonitis, intra-abdominal abscess, intestinal obstruction and infertility. Table 27.2 gives summary of various types of peritonitis.
- Other name for this type of peritonitis is abortion peritonitis.

KEY BOX 27.14

ABORTION PERITONITIS

- Instrumentation
- Puerperal sepsis
- Offensive lochia
- Pelvic peritonitis
- Infertility

SPONTANEOUS (PRIMARY) BACTERIAL PERITONITIS (SBP)

As the name suggests, in this condition, there is no demonstrable intra-abdominal disease responsible for peritonitis.

Types

1. **In infants:** It is more common in female children. Spread is by haematogenous route. The causative organisms are *Streptococcus pneumoniae.* It may follow respiratory tract or urinary tract infection (Key Box 27.15).
2. **In adults:** Male alcoholic patients are commonly affected followed by patients with chronic liver disease. Causative organisms are *E. coli, S. faecalis,* etc.
   - **Portal hypertension** increases **permeability of gut wall**, thus increasing bacterial migration. These bacteria which colonise in the small bowel reach systemic circulation **because of shunting of blood around liver** sinusoids. **Portal lymph also gets contaminated** giving rise to increased ascitic fluid.

KEY BOX 27.15

RISK FACTORS

CHILDREN

- Malnutrition
- Malignancy
- Chemotherapy
- Splenectomy

ADULTS

- Cirrhosis
- Nephrotic syndrome
- Chronic renal failure

Table 27.2 Summary of various types of peritonitis

Type	Age	Route	Exudate
Pneumococcal	3–6 years	Vagina	Odourless, sticky, turbid pus with high-grade fever
β-haemolytic streptococci	Children	Vagina	Cloudy and fibrin flakes +
Parturition	20–40 years	Vagina	Thick pus +
Tuberculous	20–40 years	Pulmonary tuberculosis	Straw-coloured fluid + tubercles +
Perforation peritonitis	20–40 years	Hollow viscus	Foul-smelling exudate +

Clinical features

- **Dull aching** pain in the abdomen with low-grade fever.
- **Rebound tenderness** is present, bowel sounds are absent or sluggish—abdominal distension
- Cirrhotic patients may develop ***coma*** with onset of primary bacterial peritonitis.
- **Septic shock** is a late feature with a high mortality rate.

Investigations

- Leukocytosis, ↓albumin, ↑prothrombin time suggests sepsis.
- **Peritoneal tap and Gram staining** of the fluid.
- **Laparoscopy** may help to rule out intra-abdominal emergencies such as perforations, etc.
- **Ultrasound** can detect nature of the liver and amount of fluid in the abdomen.
- **CT scan** when in doubt about the diagnosis.

Treatment

- Conservative treatment is followed, provided secondary bacterial peritonitis is ruled out.
- Broad-spectrum antibiotics such as aminoglycosides with 3rd generation cephalosporins is the ideal choice. Metronidazole is always added.
- Instillation of antibiotic solution into ascitic fluid to achieve a quick and high concentration.
- If laparotomy is done, peritoneal wash or toilet is given.

PERIODIC PERITONITIS

- Also called familial Mediterranean fever.
- It is of unknown aetiology
- It affects children, young adults and females.
- Presents as abdominal pain, tenderness, pyrexia and increased total WBC count.
- When in doubt, laparotomy should be done.

Some salient features (Observe 11Ps)

- **Pyrin**—a protein product from the gene—MEFVgene mutation causes this.
- **Pain** abdomen and tender abdomen
- **Pyrexia**
- **Pain** in the thorax
- **Period** of 2–3 days, it occurs and slowly remission occur.
- **Patients** are usually ***children***
- **Principally** occurs in ***Arabs, Armenians*** and ***Jews.***
- **Primarily** runs in families
- **Peritoneal** inflammation is present
- **Polymorph** leukocyte count is increased
- **Prevent** recurrence—colchicine therapy

Tumours of the peritoneum

- **Primary:** Mesothelioma is the tumour to be remembered It is more common from the pleura. In the abdomen, pelvi peritoneum is the common site.
- **Secondary** tumours are—pseudomyxoma peritonei an carcinoma peritonei.

PSEUDOMYXOMA PERITONEI

- In this condition, peritoneal cavity is filled with mucoi substance (jelly-like) brownish or yellowish.
- Mucinous cystadenoma appendix is the cause and late spreads to ovaries (Fig. 27.59). Some also believe disease starts in ovary and later spreads to other organs.
- Primary tumour is **very slow growing** and metastasis i exceptional.

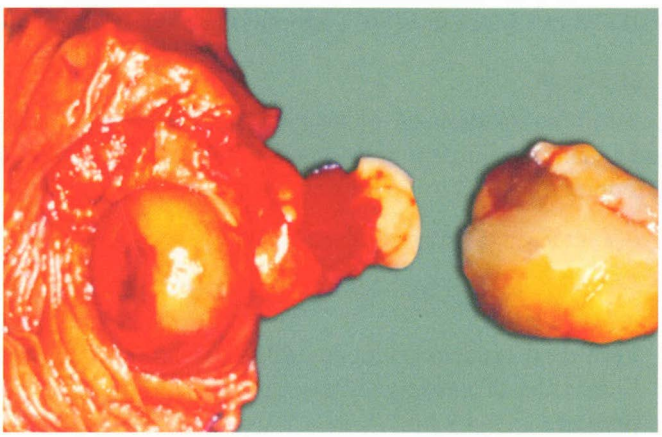


Fig. 27.59: Rupture of mucocoele of the appendix (*Courtesy:* Dr Stanley Mathew, FRCS, Professor, Department of Surgery, KMC, Manipal)

Clinical features (Key Boxes 27.16 and 27.17)

- The patients can present with slow, painless and progressive abdominal distension. No shifting dullness.
- It is more common in females.
- It can also present with features of intestinal obstruction.

Treatment

- Aggressive surgery is the main mode of treatment. Masses of tumour tissue (jelly) should be resected or scooped out. Thus debulking, bilateral oophorectomy, appendicectomy, omentectomy are done.
- Combination chemotherapy by using cisplatin and/or intraperitoneal alkylating agents have been used.

KEY BOX 27.16

SITES

- Ovary—common site
- Appendix—common site
- Intestines—not uncommon sites
- Uterus, urachus—rare sites

KEY BOX 27.17

PSEUDOMYXOMA PERITONEI

Women
Operation: Surgery is the main treatment
Mucinous cyst of appendix/ovary is the cause
Extraperitoneal spread never happens
No shifting dullness—material is jelly-like

At surgery

Masses of jelly scooped out
Appendicectomy should be done
Debulking of mass to be done
OVAriectomy, omentectomy is done
Radioactive isotopes can be instilled into peritoneal cavity
Yellow apron—omentum should be removed

Remember as **WOMEN have MAD OVARY**

- Recurrence can occur because the tumour is locally malignant.

CARCINOMA PERITONEI

- This name is applied to an advanced stage of intra-abdominal malignancy involving the entire peritoneal cavity.
- The causes are shown in Key Box 27.18.

KEY BOX 27.18

COMMON CAUSES

Carcinoma of the:
- Stomach, colon, pancreas
- Breast, ovary

Features at laparotomy

- Multiple firm to hard nodules on the visceral and parietal peritoneum.
- Dense adhesions between the intestinal loops and other viscera.
- Plaques on the intestinal surface
- Widespread secondaries in the liver
- Entire omentum will be studded with hard nodules. It is called omentum cake (Fig. 27.60).
- Ascites: Straw-coloured or haemorrhagic
- Greater omentum being the **policeman** and a great drain pipe of the abdomen with rich lymphatics, is studded with nodules.
- Low protein ascites is more vulnerable for risk of developing peritonitis. **Incidence of peritonitis in malignant ascites cases is low because of increased immunoglobulin levels in malignant ascites and increased opsonic activity.**

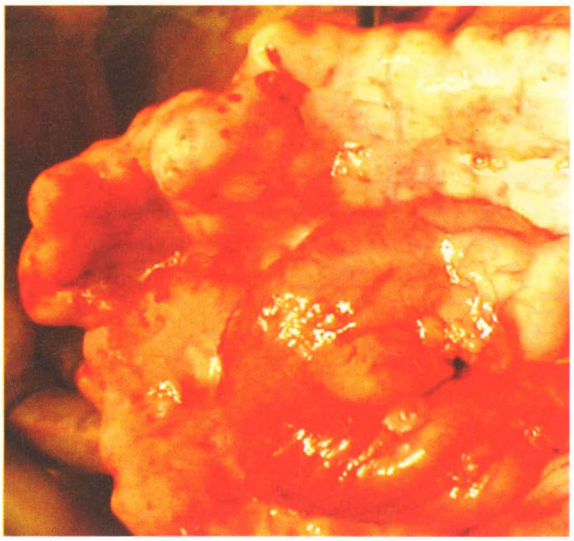


Fig. 27.60: Omental cake in colorectal cancer

Differential diagnosis

- The most common differential diagnosis is tuberculosis (peritoneal). These nodules are firm and greyish.
- Acute pancreatitis with fat necrosis: This is due to calcium soap. They are yellow and soft.
- Ruptured hydatid cyst
- Lymphomatous nodules

Treatment

1. Radioactive gold (Au^{198}) instillation into peritoneal cavity.
2. Tamoxifen is useful for ascites due to carcinoma of the breast.

GRANULOMATOUS PERITONITIS

- Talc, gauze, starch, etc. are causative factors.
- It occurs many weeks after surgery.
- Low-grade fever, weight loss, distension, crampy abdominal pain are the features.
- Laparoscopy is the key investigation, can visualise granuloma and biopsy can be taken. Also, fluid can be aspirated and sent for histopathology. High concentrations of lymphocytes are present in both.
- Symptomatic treatment—sinus, fistula needs to be treated.
- Intravenous prednisolone followed by oral prednisolone for 2–3 weeks is given.

Prevention

- Cleaning the gloves by wiping before handling bowel, prevents many cases of granulomatous peritonitis.
- In the initial surgery all the foreign bodies including ova, cysts of parasites, ascariasis, ingested food particles have to be removed.

THE OMENTUM

Surgically important diseases in the omentum are:

1. **Tuberculous peritonitis:** Here omentum is involved. It becomes nodular because it is studded with tubercles. Classically seen in children who are brought with abdominal distension. On palpation, omentum is felt as granular mass in the upper abdomen which moves with respiration. Laparoscopy, biopsy and antitubercular treatment is given in Fig. 27.61.
2. **Metastasis:** Carcinoma stomach, colon, pancreas commonly result in metastasis and they give rise to omental cake (Fig. 27.62).
3. **Tumour (cyst):** Omentum is the site of omental cyst which is a lymphatic cyst (Fig. 27.63). It is a slow-growing, painless swelling in the upper abdomen. On examination, the patient is a child or an adult, with a smooth, firm mass in epigastrium which moves with respiration. Excision of the lymphatic cyst is the treatment. Massive cyst can be confused for ascites.

Fig. 27.61: Rolled up omentum in tuberculosis

Fig. 27.62: Omental cake in metastasis

Fig. 27.63: Omental cyst (lymphatic)

Fig. 27.64: Omentum in non-Hodgkin's lymphoma (NHL)

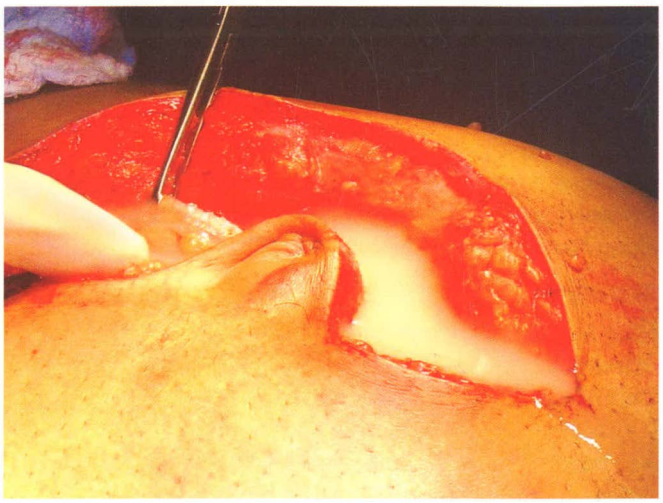


Fig. 27.65: Pus-like milky fluid once abdomen is opened (NHL)

4. **Torsion:** It is a rare surgical emergency wherein torsion of omentum occurs due to old **adhesions** or it is primary due to lengthy mobile omentum. It produces symptoms/signs similar to that of appendicitis. Laparotomy and excision of gangrenous omentum have to be done.
5. Non-Hodgkin's lymphoma can affect ometum and can give rise to granularity (Figs 27.64 and 27.65).

Complications: Haemorrhage into the cyst.

MESENTERY

- **Anatomy:** Given in page 666
- Mesenteric tear has been discussed in page 667
- Mesenteric lymphadenitis has been discussed in page 672

MISTY MESENTERY

- It means increase in the mesenteric fat and it is a finding in the multi-detector CT scan.
- **A few pathological conditions which may give rise to this entity are:** Acute pancreatitis, retroperitoneal haemorrhage, malignancies.
- Once the source is treated, the findings may reduce or may disappear.
- Hence, follow-up CT scans are required.
- **Weber-Christian disease:** Lipodystrophy and mesenteric lipogranuloma are the features. In this condition, it should be called mesenteric panniculitis. Findings include inflammation of the mesentery, fibrosis, shortening and ischaemia.It is difficult to treat.

MESENTERIC CYST

- These are congenital cysts, enterogenous or chylolymphatic. It manifests in young children or during adolescence. Typically, the cyst is located in the umbilical region which moves at right angles to the direction of mesentery (Fig. 27.66).

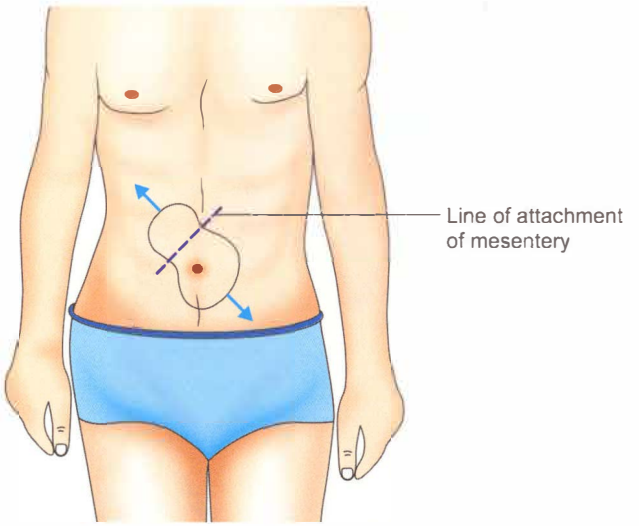


Fig. 27.66: Diagrammatic representation of mesenteric cyst and its movements at right angles to the direction of mesentery

Types of mesenteric cysts (Figs 27.67 to 27.70, Key Box 27.19)

A. Chylolymphatic cyst is a lymphatic cyst arising from mesentery of ileum. It is a thin-walled cyst with clear fluid or chyle. It has a separate blood supply. Hence, enucleation is the treatment without sacrificing the bowel.

B. Enterogenous cyst is a duplication cyst from the intestine or due to diverticulum of the mesenteric border of the intestine. It is thick walled and contains mucus. This cyst is treated by excision of cyst with bowel segment because both share the same blood supply.

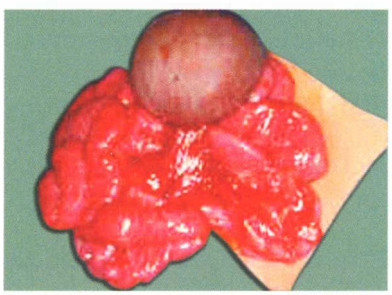


Fig. 27.67: Enterogenous cyst at surgery

Tillaux's Triad

1. Fluctuant swelling near the umbilicus.
2. Movement perpendicular to the line of mesentery.
3. It is dull surrounded by a zone of resonance and traversed by band of resonance.

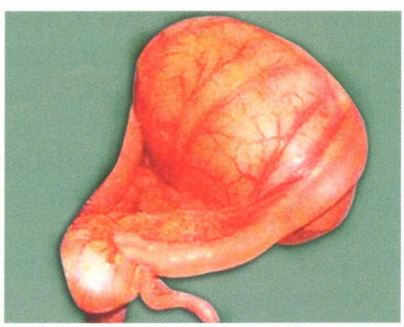


Fig. 27.68: Mesenteric cyst excised in toto

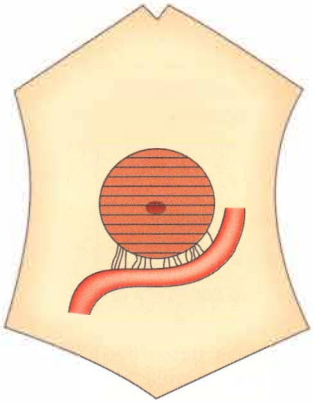


Fig. 27.69: Chylolymphatic cyst—it can be excised without resection of the bowel

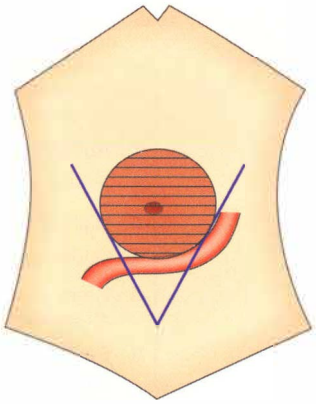


Fig. 27.70: Enterogenous cyst—it requires excision of intestine along with the cyst

Complications

- Torsion of the cyst resulting in acute abdominal pain.
- Rupture of the cyst due to trauma
- Haemorrhage into the cyst

KEY BOX 27.19

MESENTERIC CYST—TYPES

- Chylolymphatic cyst
- Enterogenous cyst
- Urogenital remnant
- Teratomatous dermoid cyst

RETROPERITONEUM

ANATOMY

Retroperitoneal space: The retroperitoneal space lies between the peritoneum and the posterior parietal wall of the abdominal cavity and extends from the diaphragm to the pelvic floor.
Superiorly—12th thoracic vertebra and lateral lumbocostal arch.
Inferiorly—base of the sacrum, iliac crest, and iliolumbar ligament.
Anteriorly—posterior parietal peritoneum
Posteriorly—fascia overlying the quadratus lumborum and psoas major muscles.

Important anatomical organs (Fig. 27.71)

1. **Urinary:** Adrenal glands, kidneys, ureter, bladder
2. **Circulatory:** Aorta, inferior vena cava
3. **Digestive:** Oesophagus (thoracic part), rectum (part of middle third and lower third is extraperitoneal).
4. The head, neck, and body of the pancreas, the duodenum, except for the proximal first segment, ascending and descending portions of the colon.

Posterior abdominal wall includes the study of the following structures

1 Abdominal aorta
2 Inferior vena cava
3 Right and left kidneys with ureters
4 Duodenum on the right side
5 Head of pancreas in the concavity of duodenum
6 Body and tail of pancreas across the posterior abdominal wall towards the left kidney

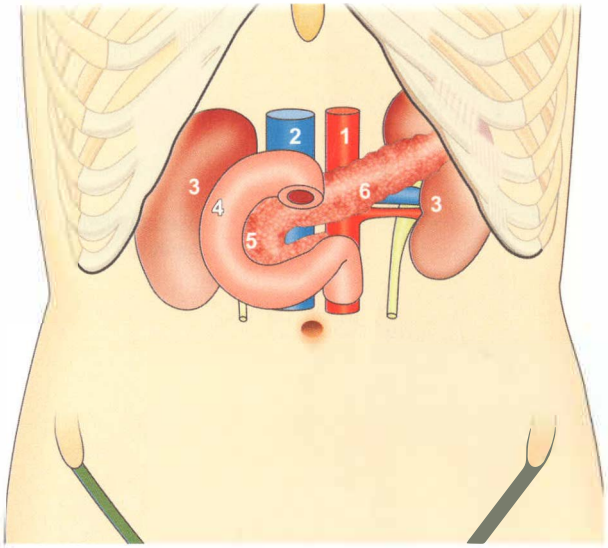


Fig. 27.71: Anatomical organs in the retroperitoneal space

IDIOPATHIC RETROPERITONEAL FIBROSIS—ORMOND'S DISEASE

- This is one of a group of fibromatosis (other being Dupuytren's contracture and Peyronie's disease).
- Other types of fibrosis such as mediastinal fibrosis, sclerosing cholangitis may also be associated features.
- Riedel's thyroiditis may be associated with this condition
- As collagen encases ureters, they present with ureteric obstructions, requiring ureteric stenting.
- Steroid therapy is the treatment of choice
- Tamoxifen may help
- It can present with lower back pain, renal failure, hypertension, deep vein thrombosis and other obstructive features.

See Key Box 27.20 and clinical notes.

KEY BOX 27.20

IDIOPATHIC RETROPERITONEAL FIBROSIS—CAUSES

Idiopathic—Ormond's disease
Drugs—chemotherapy, methysergide, β-adrenoreceptor antagonists
Irritation—blood, urine, bowel contents
Others—autoimmune
Peritoneal disease—secondaries
Aortic aneurysm—inflammatory type
Trauma
Hereditary/familial
Inflammation—chronic
Carcinoid

Remember as **IDIOPATHIC**

CLINICAL NOTES

- *One day I was urgently called to assist the urosurgeon who had opened the abdomen for ureterolysis. Following was the clinical picture.*
- *A 76-year-old lady was admitted under urology for the diagnosis of renal failure. An ultrasound abdomen showed bilateral hydronephrosis. She was diagnosed to have idiopathic retroperitoneal fibrosis and posted for ureterolysis. To my surprise, when I explored, the patient had carcinoma colon in three places (synchronous), carcinoma caecum infiltrating right ureter, carcinoma sigmoid colon infiltrating left ureter and carcinoma transverse colon. She underwent total colectomy with ileorectal anastomosis. Ureteric reimplantations were done into bladder. She lived for 3 years and then succumbed due to liver metastasis.*

RETROPERITONEAL CYST

- Very often, it is a painless, smooth, firm enlargement.
- May have minor degree of mobility.
- These cysts are either lymphatic cysts or derived from the remnant of Wolffian duct. A few of them are teratomatous dermoids.
- Benign retroperitoneal cysts are usually mesothelial or mesonephric in origin; rarely, rupture of the biliary tree can result in bile-filled cysts.
- **Gross:** These cysts are not connected to the kidney or adrenal; usually filled with clear or straw-coloured fluid.
- **Histology:** These cystic structures may be lined by mesothelial, enteric (glandular), or columnar epithelium.

- They can be unilocular or multilocular.
- CT scan is required to differentiate it from hydronephrosis.
- Excision is the treatment.

RETROPERITONEAL ABSCESS

Causes

1. Renal source: Pyonephric abscess
2. **Spine: Tuberculosis (details below)**
3. Haematoma: Fracture spine/pelvis
4. Acute pancreatitis (on right side)
5. Retrocaecal appendicitis (on right side)
6. Sigmoid diverticulitis

Diagnosis

- Ultrasound or CT scan-guided aspiration
- Culture of the pus and antibiotic sensitivity.

Treatment

Aspiration, appropriate antibiotics and open drainage.

PSOAS ABSCESS

Three types have been recognised

1. **Primary psoas abscess:** It is caused by haematogenous spread of *Staphylococcus aureus*. Source may be occult—tonsils, middle ear, etc. More common in children and young adults. Poor nutrition, may be a contributing factor—It is **monomicrobial**.
2. **Secondary psoas abscess:** Secondary to intestinal perforation, e.g. Crohn's disease. It is **polymicrobial.** Other causes are given below.

Clinical features

Fever, flank pain, flexion of the hip joint are triad of psoas abscess. Pain on extension confirms the diagnosis.

Management

- CT scan is the diagnostic test. Gas bubbles are diagnostic of an abscess.
- Treatment include percutaneous catheter drainage, treatment of the source of infection with antibiotics.
- If necrotic tissue does not drain well or if patient is not improving, open drainage should be done.

3. **Tuberculous spine:** Lower thoracic (T10) and upper lumbar spine are commonly affected (Fig. 27.72).

Clinical features

- Pain in the back (localised to lesion) or referred pain if there is a collapse.
- Evening pyrexia
- Protective muscular spasm, especially of sacrospinalis.
- Collapse of the anterior portion of vertebral body results in angular deformity—gibbus.

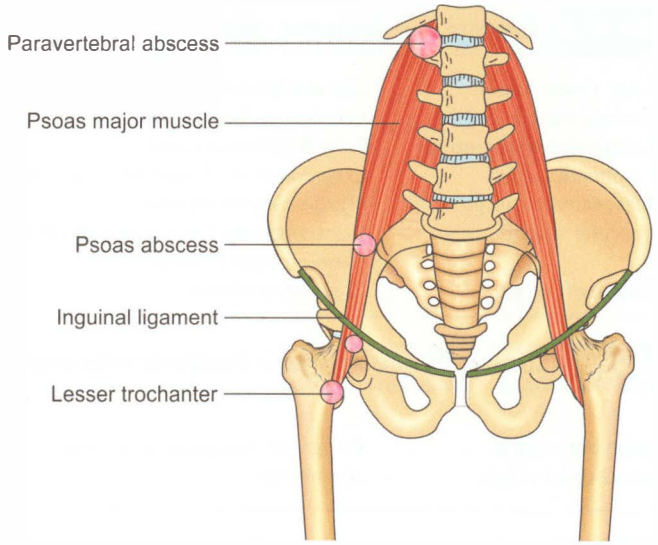


Fig. 27.72: Various sites of tubercular cold abscess arising from tuberculosis of the spine

Route of psoas abscess

- Pus enters psoas sheath and tracks downwards and causes mass in the iliac fossa.
- From here, it traverses beneath inguinal ligament.
- If untreated, it collects in the subcutaneous plane.

Investigations

- **Chest X-ray, ESR, sputum AFB**
- **Spine X-ray**—AP and lateral view. The earliest sign is a decrease in the intervertebral space.
- **MRI** can detect spine lesion, cold abscess
- **CT/MR-guided aspiration of pus**/biopsy to prove histological diagnosis.

Treatment

- Cold abscess—aspiration followed by antituberculous treatment.
- Unstable/collapse spine—costotransversectomy—lateral thoracotomy.

RETROPERITONEAL TUMOUR

Definition

- The term retroperitoneal tumour (**RPT**) is usually confined to primary tumours arising in other tissues in this region, e.g. muscle, fat, lymph nodes, nerves (Tables 27.3 to 27.5). (**Jean Lobstein**, French pathologist and surgeon in 1829—coined the term **retroperitoneal tumour.**)
- Other tissues refer to—tumours of retroperitoneal organs such as kidneys, ureters, pancreas and adrenals are conventionally not included in retroperitoneal tumours.

Table 27.3 Tumours of mesodermal origin (75%)

Fatty tissue origin	*Lipoma/liposarcoma*
Smooth muscle origin	*Leiomyoma/leiomyosarcoma*
Connective tissue origin	*Fibroma/fibrosarcoma*
Lymphatic origin	*Lymphangioma/lymphangiosarcoma*
Mesenchymal origin	*Myxoma/myxosarcoma*
Vascular origin	*Haemangioma/haemangiosarcoma/ haemangiopericytoma*
Unknown origin	*Xanthogranuloma*

Table 27.4 Tumours of notochordal or embryonic rests origin

Embryonic origin	*Benign or malignant teratomas*
Notochordal origin	*chordomas*

Table 27.5 Tumours of neurogenic origin (25%)

Germ cell origin	*Benign or malignant teratomas*
Sympathetic origin	*Chordomas*
Adrenochromaffin origin	*Cortical carcinoma/ paraganglioma/ pheocromocytoma*

Introduction

- Uncommon (0.2–0.6% of all tumours)
- **Malignant** in 80–85% of cases (of these, 35% are sarcomas).
- **Sex:** No differences
- **Age:** Most occur between the sixth and seventh decade of life.
- Because of their location, these lesions usually demonstrate **indolent growth** and present as relatively large lesions.
- Their proximity to vital structures (especially vascular) makes **resection difficult**.

Aetiology

1. Idiopathic: The actual cause is not known
2. History of radiation: Accidental or given for lymphoma.
3. Exposure to vinyl chloride/thorium dioxide
4. **Familial disorders:** Gardner's syndrome, familial neuroblastoma, neurofibromatosis, Li-Fraumeni syndrome.

Symptoms of RPT

- **Abdominal mass (80%):** Slow growing painless mass is the most common presentation. Typically confined to one side rather than centre.
- **Nausea, vomiting** and weight loss in about 20 to 30% of patients.
- **Compressive symptoms:** Abdominal pain, constipation, recent haemorrhoids, haematochezia can occur. Back pain and sciatica (30%) are common and are confused for spine pathology.

Unilateral lower extremity oedema and pressure symptom including secondary varicosities are common.

Acute urine retention, dysuria and increased frequency cai occur due to compression on urinary bladder

- **Paraneoplastic syndrome** such as intermittent hypo glycaemia can occur in liposarcoma/fibrosarcoma anc catecholamine excess in paragangliomas (Key Box 27.21)

KEY BOX 27.21

WHEN TO SUSPECT RETROPERITONEAL SARCOMA

- Recent finding of large abdominal mass
- Recent lower limb swelling
- Recent varicosities
- Recent varicocele
- Recent haemorrhoids
- Recent loss of weight

Signs of RPT

- Usually large abdominal mass, firm to hard, irregular.
- Nonmobile, restricted mobility
- Not moving with respiration
- Does not fall forward (knee elbow position)
- Resonant tone on percussion—due to bowel anteriorly.
- Transmitted pulsations may be felt.

Differential diagnosis

- Lymphoma
- Germ cell tumours
- GIST
- Metastatic testicular cancers

Please note: When you suspect anyone of these as differential diagnosis a CT-guided biopsy may be required which alters the management.

Investigations

Preoperative planning to assess the extent of the disease is essential because successful surgery is defined by complete excision of the mass with adequate margins of normal tissue.

1. CBP
2. **LFT:** Increase alkaline phosphatase may indicate secondaries.
3. RFT: Compressive uropathy with high urea and creatinine.
4. Tumour markers: AFP, beta-HCG—germ cell tumour.
5. **LDH in lymphoma/GCT:** LDH levels indicate tumour burden and growth rate.
6. **CT Scan** (Key Box 27.22)
   - Delineate the anatomic limits of the lesion.
   - Vascular involvement—vena cava, aorta, renal vessels.
   - Assess the integrity and function of adjacent organs—renal function is one of the important advantage of CT scan.
   - Visceral metastases +/–. If present, it is inoperable but still worth trying a resection after giving neoadjuvant therapy.
   - Para-aortic, iliac, mesenteric lymphadenopathy.
   - Axial skeleton and renal involvement (Key Box 27.22).

KEY BOX 27.22

CT SCAN: TYPICAL FINDINGS

- **Lipoma:** Homogenous fatty density
- **Malignant fibrous histiocytoma:** Calcifications
- **Neurofibroma:** Homogenous low density
- **Teratoma:** Mixed components
- **Paraganglioma:** Para-aortic location
- **Neuroblastoma:** Calcified tumour, usually in children
- **Leiomyosarcoma:** Large areas of necrosis
- **Liposarcoma:** Heterogenous fatty density
- **Hemangiopericytoma:** Hypervascularity

7. **CT-guided core biopsy**
   - Reserved for cases in which a diagnosis will change therapy, such as the need for neoadjuvant chemotherapy for
     1. GIST—imatinib mesylate
     2. Germ cell tumours
     3. Lymphomas
8. **Laparoscopic biopsy/retroperitoneoscopy**
   - Equivocal history and diagnostic dilemma
   - Unusual appearance of the mass
   - Unresectable tumour
   - Distant metastasis

Treatment: Surgery is the main modality and the most effective modality of the treatment. It can be curative if R-0 resection is achieved. Chemotherapy, radiotherapy are complementary to surgery. A few cases are also managed by neoadjuvant therapy.

I. Surgery

Principles: Extirpative surgery is the principal and most effective form of therapy for primary retroperitoneal tumours. **Tumour histology, tumour size, or patient age** *not significant factors in survival* in multiple-variable analysis. Therefore carefully planned and skillfully executed surgical therapy is critical for any chance at long-term success.

Adjuvant therapy

High local recurrence rate and eventual mortality from this disease has prompted the exploration of adjuvant therapeutic modalities. **Postoperative radiation increased toxicity to surrounding structures.**

II. Radiotherapy

- Radiotherapy: Two types

1. EBRT—external beam radiotherapy
2. Brachytherapy for retroperitoneal leiomyosarcoma.

III. Chemotherapy

- Doxorubicin is the foundation for chemotherapy in advanced sarcoma.

TEN COMMANDMENTS

1. **Complete R-0 resection should be the aim**—*single most important positive predictive feature.*
2. Generous incision should be given to get into good access to the tumour
3. Intra-abdominal adhesions should be released to separate the tumour. Should use of **sharp, curved Mayo scissors** for meticulous dissection.
4. Should never do enterotomy (accidental) because an enterotomy and subsequent fistula formation can cause major morbidity.
5. Should preserve all important vessels.
6. Should preserve important organs such as kidney, ureter colon, etc.
7. Should resect the organs if found infiltrated and if R-0 resection is possible.
8. Should do a **centripetal dissection.** It allows one to dissect those areas that are amenable to dissection.
9. **Should use surgical clips,** should be placed to mark the periphery of surgical field and other relevant structures to help guide potential future radiation therapy.
10. Both radiotherapy and chemotherapy are given.

- **MAID regimen:** Mesna, adriamycin (doxorubicin), ifosfamide, and dacarbazine has been successful in neoadjuvant programmes for sarcomas of the extremities compared with historical controls. Yet less data are available with regard to other soft tissue sites. Neoadjuvant therapy for retroperitoneal sarcoma.

Advantages

- The viscera are often displaced by the tumour volume and a lack of surgical adhesions further reduces dose to the bowel.
- Effective radiation dose is lower in the preoperative setting.

DIFFERENTIAL DIAGNOSIS OF RETROPERITONEAL SARCOMA

These are pathological variants. It is difficult to consider them as a diagnosis on clinical grounds. However, more common ones have to be considered first such as liposarcoma. Patients with von Recklinghausen's disease may have neural tumours. A few differential diagnosis are given below.

LIPOSARCOMA

Most common of primary RPT, 20% from retroperitoneum.

- **Histological types**
  1. Well-differentiated liposarcoma (low grade)
  2. Myxoid/round cell liposarcoma (50%–most common)
  3. Pleomorphic liposarcoma (10–15%—high grade)
  4. Dedifferentiated liposarcoma—the rate of metastasis depends on the degree of tumour differentiation, with nearly 90% of poorly differentiated tumours metastasizing.

- **Pathology:** The key feature of a liposarcoma is the lipoblast, which is essentially an immature fat cell. **LIPOBLAST** have multiple fat vacuole which compress the nucleus, creating a scalloped appearance.

LEIOMYOSARCOMA

- 50% from retroperitoneum—Female: Male—2:1
- Site of origin (soft tissue, vascular or superficial), although many of the soft tissue lesions are believed to originate from smaller blood vessels.
- Immunohistochemistry stain for smooth muscle myosin, vimentin, and actin and less often for desmin. (*Leiomyomas stain positive for desmin, which separates them from their malignant counterpart.)*
- They stain negative for S-100.

MFH (MALIGNANT FIBROUS HISTIOCYTOMA)

- Less common in the retroperitoneum.
- Derived from fibroblast differentiation *(previously defined as a sarcoma of primary histiocytic origin)*
- Storiform-pleomorphic (40–60%) and myxoid type (25%) are subtypes. Other types being giant cell type and inflammatory.

RETROPERITONEAL TERATOMA

- These are the tumours arising from totipotential cells. Thus, they can have ectoderm, mesoderm or endoderm elements.
- 10% of all primary RPT
- Rare in adults because of its congenital nature.
- Solid teratoma malignant (likely).
- Malignant mature cystic teratomas (0.2 to 2% of cases) have the potential to metastasise to sites such as the retroperitoneal lymph nodes and lung parenchyma.

RHABDOMYOSARCOMA

- 6% in retroperitoneum
- More common in children
- Sporadic—most common
  - Genetic risk factor—10–33%: Li-Fraumeri syndrome, neurofibromatosis.

SCHWANNOMA

- Majority has mutations in *NF2* gene.
- Majority are sporadic tumours.
- A minority (10%) are associated with syndromes such as neurofibromatosis type 2, schwannomatosis and multiple meningiomas.
- Hallmark of schwannoma is alternating areas of cellular (Antoni A) and hypocellular (Antoni B) areas.
- Retroperitoneal tumours are larger and often show degenerative changes such as cystic change, haemorrhage and calcifications.
- Immunohistochemistry: Diffuse S-100+ is characteristic.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated with flowcharts and coloured pictures.
- More details on mesentery and retroperitoneum are added.
- Retroperitoneal mass/sarcoma and abdominal compartment syndrome are discussed in more detail.
- Misty mesentery valentino syndrome and mesenteric cyst have been added.

MULTIPLE CHOICE QUESTIONS

1. One need not close peritoneal layer after laparotomy because:
A. The peritoneum can get stuck to the bowel
B. Flattened mesothelial cells heal within a few hours
C. The peritoneum tears when closure is attempted
D. It is very painful postoperatively

2. Peritoneum can be used for dialysis because:
A. It is close to kidney
B. It is faster than haemodialysis
C. Capillaries and lymphatics between two layers of peritoneum help in absorption and exudation
D. It covers entire abdomen

3. Which of the following is an example of primary peritonitis?
A. Tuberculous peritonitis
B. Perforation peritonitis
C. Postoperative peritonitis
D. Parturition peritonitis

4. Which of the following organisms are most commonly involved in secondary peritonitis?
A. Enterococci
B. Streptococci
C. Staphylococci
D. Pneumococci

5. The following are the typical features of acute generalised peritonitis *except:*
A. Abdominal pain
B. Persistent vomiting
C. Bradycardia
D. High-grade fever with chills

6. Abdominal tap is done in peritonitis for all of the following roles *except*:
A. Aspiration of blood to indicate haemoperitoneum
B. Aspiration of pus indicating infection with gram-negative bacteria
C. Aspiration of bile indicating biliary peritonitis
D. Aspiration of urine indicating ureterocoele

7. The following suture material is best suited for closure of bowel perforation:
A. Silk B. Catgut
C. Nylon D. Thread

8. History of discharge per rectum for the first time in a patient who is recovering from peritonitis suggests:
A. Anal prolapse B. Pelvic abscess
C. Proctitis D. Colitis

9. What forms the anterior relationship of Rutherford Morrison's space?
A. Liver B. Kidney
C. Diaphragm D. Duodenum

10. Subphrenic abscess is common on the right side because of the following reasons *except*:
A. Majority of the diseases affect right side
B. Right lung is larger
C. Left paracolic gutter is narrow and colophrenic ligament is present on the left side
D. Right paracolic gutter is large and colophrenic ligament is absent on the right side

11. Indications for open drainage of subphrenic abscess include the following *except:*
A. Persistent fistula discharging pus
B. Thick viscid pus
C. Abscess very close to IVC/diaphragm
D. Single loculus

12. Intra-abdominal pressure exceeds _____ cm H_2O in abdominal compartment syndrome.
A. 15 B. 25
C. 35 D. 45

13. Following are features of tuberculous peritonitis *except*:
A. Tubercles over peritoneal surface
B. Encysted form
C. Can be a military form
D. Transudate

14. Nonoperative treatment for peritonitis may be followed in the following *except:*
A. Moribund patients
B. Sealed perforation
C. Localised peritonitis
D. Generalised peritonitis

15. The following catheters are commonly used for percutaneous drainage of subphrenic abscess:
A. Pigtail catheter B. Trocar catheter
C. Sump catheter D. Foley's catheter

16. More reliable sign of peritonitis is:
A. Cough tenderness
B. Tenderness on pressure
C. Rebound tenderness
D. Guarding

17. Presence of sunken eyes, pale and pinched face, dry cracked tongue, cold perspiration and cyanosis are all typical features of:
A. Hippocratic facies B. Gargoyle facies
C. Marshall hall facies D. Mask like facies

18. Following are risk factors for spontaneous bacterial peritonitis *except*:
A. Cirrhosis
B. Nephrotic syndrome
C. Chronic renal failure
D. Carcinoma stomach

19. Which of the following is true for pneumococcal peritonitis?
A. Common in young boys
B. Age is around 15 years
C. Peritoneal fluid is transudate
E. It is typically odourless

20. Following are about pseudomyxoma peritonei *except*:
A. Common in women
B. Ovary is the main source
C. Surgery cannot cure the disease
D. Chemotherapy is also used

ANSWERS

1 B	2 C	3 A	4 A	5 C	6 D	7 A	8 B	9 D	10 B
11 D	12 C	13 D	14 D	15 A	16 C	17 A	18 D	19 D	20 B

28

Small Intestine

- Anatomy
- Physiology
- Abdominal tuberculosis
- Tuberculous peritonitis
- Glandular tuberculosis
- Intestinal tuberculosis
- Inflammatory bowel diseases
- Ileostomy
- Surgical complications of enteric fever
- Intestinal amoebiasis
- Radiation enteropathy
- Peutz-Jegher syndrome
- GIST
- Carcinoid tumour
- Short gut syndrome
- Diverticula
- Intestinal fistula
- What is new?/Recent advances

Introduction

Truly speaking small intestines extends from pylorus to ileo-caecal junction. However, for all practical purposes it is discussed as starting from duodenojejunal flexure till caecum. Small intestines play an important role not only in the transfer of food contents distally but in the digestion, absorption and secretion of the contents. Being the central portion of the GI tract with long length, many diseases affect the intestine. Surgically important topics have been covered in this chapter.

ANATOMY

- Small intestine consists of proximal 2/5 jejunum and distal 3/5 ileum. It is about 6 metres in length.
- Small intestine starts at duodenojejunal flexure just to the **left of the inferior mesenteric vein**.

  Surgical importance: To identify the first (short) loop of jejunum for gastrojejunostomy.
- Small intestine ends at ileocaecal junction.

In cases of intestinal obstruction, trace up to ileocaecal junction. If caecum is distended, it is a case of large bowel obstruction. If caecum is collapsed, it is a case of small bowel obstruction.

- Jejunum resides in the left side of the peritoneal cavity and ileum on the right side.
- Differences between jejunum and ileum have been given in Key Box 28.1.

KEY BOX 28.1

DIFFERENCES BETWEEN JEJUNUM AND ILEUM

	Jejunum	Ileum
• **Length**	2/5	3/5
• **Diameter**	Wider (2–4 cm)	Less (2–3 cm)
• **Wall**	Thick and double (mucous membrane can be felt)	Thin
• **Colour**	Deep red	Pale pink
• **Peyer's patch**	Very, very less	More
• **Blood supply**	Long and few vasa rectae (1 or 2)	Short and numerous (5 or 6)
• **Mesentery**	Transparent, less fat	More fat

Blood Supply (Fig. 28.1)

- Superior mesenteric artery is the **artery of the midgut** which supplies the entire midgut (entire small intestine). Jejunal arteries are end-arteries.
- Mesenteric border of the intestine gets more blood supply when compared to anti-mesenteric border. Hence in cases of diminished blood supply, anti-mesenteric border become ischaemic first.
- Venous drainage is through superior mesenteric vein.

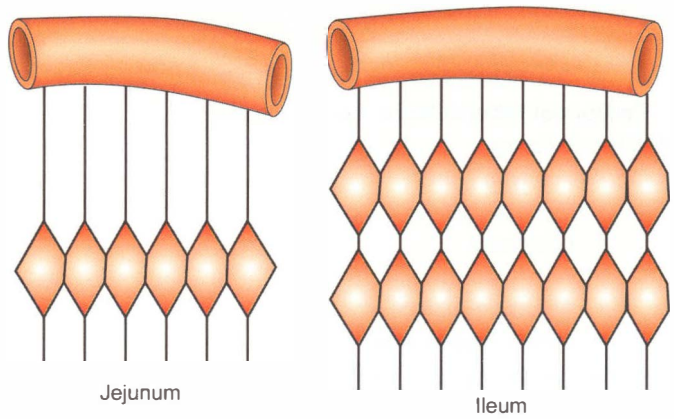


Fig. 28.1: Blood supply—jejunum and ileum

Mesentery

- It is a fan-shaped fold of peritoneum which attaches jejunum and ileum to posterior abdominal wall.
- Blood vessels and lymphatics course in between the folds of peritoneum.
- It extends from the left of duodenojejunal flexure (left of L2) vertebra to the right sacroiliac joint, thus fixing the ileocaecal junction there.
- The importance of direction of the mesentery is appreciated in the following examples:
  - **A. Mesenteric cyst** moves at right angles to the direction of the mesentery.
  - **B. Mesenteric lymph nodes** can be clinically palpable as a nodular or a smooth mass.
  - **C. Horizontal tear** in the mesentery causes more gangrene of the bowel than vertical tear (Fig. 28.2).
- **Structures crossed by mesentery:** Duodenum, aorta, inferior vena cava, right ureter, right psoas major and right gonadal vein.

Fig. 28.2: Observe blood supply of the mesentery and it is but natural that a horizontal tear causes more gangrene

Microscopic Anatomy

- Basic unit of small bowel mucosa is the villus, which is a finger-like projection. Each villus is covered with tall columnar epithelium.
- Goblet cells, Paneth cells and endocrine cells are seen in the crypts.

Goblet cells are mature mucous cells. Endocrine cells (enterochromaffin cells) have cytoplasmic granules which secrete 5-hydroxytryptamine, neurotensin, glucagon and motilin. Importantly, mucosal cell mediated immunity is brought by mucosal T lymphocytes.

FUNCTIONS – PHYSIOLOGY

- **Motility:** Two types of muscle contractions occur—one which does not propagate—it exposes the food contents to the absorptive surface for a longer time by causing segmentation thus better absorption of the food. Another type is peristaltic which propagates the food contents. Control of peristalsis is done by myenteric plexus. Time taken by the solid food contents to reach from mouth to colon is about 4 hours (Key Box 28.2).
- **Absorption and digestion:** Except calcium and iron almost everything is absorbed in the small intestines. To give a few examples: out of 6–10 litres of water, almost 80% water is absorbed in the small intestine and only 10–20% is discharged into colon. Thus in cases of terminal ileal obstruction, about 8–10 litres of fluid accumulate resulting in gross distension of the abdomen and dehydration. Carbohydrates and fat are mainly absorbed in duodenum and proximal jejunum. Proteins require pancreatic enzymes. Hence, they are broken down in the jejunum into amino acids and peptides. Conjugated bile acids are absorbed in the terminal ileum wherein enterohepatic circulation takes place and again they are secreted in the bile. Thus in ileal resections or diseases like Crohn's disease, more amount of bile acids enter colon resulting in diarrhoea due to increased secretion of water and electrolytes.

KEY BOX 28.2

FUNCTIONS OF THE SMALL INTESTINE

- Digestion and absorption
- Synthesis of lipoproteins
- Secretion of regulatory peptides
  - Secretin
  - Cholecystokinin
  - Somatostatin
  - VIP
- Immune function: Production of immunoglobulins (IgA). The **B** cells and **T** cells help in phagocytosis and secretion of cytokines.

Diseases of the small intestines

Following are the various diseases of the small intestines described in the texts.

- **Abdominal tuberculosis** which includes intestinal, mesenteric and peritoneal tuberculosis. We should realise that all three types are very much connected to each other in one way or other and hence are discussed together.
- **Inflammatory bowel disease** includes ulcerative colitis and Crohn's, both are important health problems in the Western world. Other inflammatory conditions include enteric fever and its manifestations, intestinal amoebiasis, HIV associated enteropathy and radiation-induced enteropathy.
- **Tumours** include benign and malignant tumours including carcinoid tumours.
- Small intestinal obstruction, Meckel's diverticulum, superior mesenteric vascular occlusion and necrotising enterocolitis have been discussed in Chapter 30 on intestinal obstruction.

ABDOMINAL TUBERCULOSIS (TB)

Introduction

Abdominal tuberculosis is a common extrapulmonary manifestation of tuberculosis. Incidence in the West has increased due to immigrant population and increased incidence of HIV infections. In India, extrapulmonary TB is also showing re-emergence due to incomplete treatment and occurrence of multidrug-resistant strains. Hence, it is a major health problem.

Definition

- The term abdominal tuberculosis includes tuberculous infection of gastrointestinal tract, mesenteric lymph nodes, peritoneum, omentum and solid organs related to gastrointestinal tract such as liver and spleen.

Classification (Key Box 28.3)

The commonly encountered four forms of tuberculosis are given below:

1. Tuberculous peritonitis
2. Tuberculous mesenteric lymphadenitis—glandular tuberculosis
3. Intestinal tuberculosis
4. Tuberculosis of solid viscera such as liver and spleen.

Routes of spread of infection and pathogenesis

1. Intestinal tuberculosis is caused by *Mycobacterium tuberculosis* from **swallowed sputum** (pulmonary tuberculosis) or milk (milk-borne infection—*Mycobacterium bovis*). From intestinal tuberculosis, mesenteric nodes get involved and later, the peritoneum can get involved.
2. **Blood spread:** Infection from pulmonary tuberculosis can spread through blood during bacteraemic phase.
3. **Lymphatic spread** from tuberculosis of intestines.

KEY BOX 28.3

VARIOUS FORMS OF ABDOMINAL TB

I. **Peritoneal tuberculosis**: Acute, chronic
A. Chronic forms
  1. Ascitic type (wet)
    - Generalised
    - Localised
  2. Fibrous type (dry)
    - Adhesive, plastic
    - Miliary nodule type
B. Tuberculosis of peritoneal folds
  1. Mesenteric adenitis
  2. Mesenteric cysts/abscesses
  3. Bowel adhesion

II. **Gastrointestinal**
  1. Ulcerative
  2. Hyperplastic
  3. Sclerotic/plastic

III. **TB of solid viscera**
  1. Liver
  2. Spleen

4. **Genitourinary tuberculosis:** From here, cephalad spread occurs and thus, peritoneum gets affected.
5. **From bile:** Granuloma in liver. Bacilli are excreted in bile. Pathology and pathogenesis (Fig. 28.3).

Investigations

It is important to realise that there are so many investigations for abdominal tuberculosis. **Although it is essential to have a theoretical knowledge about all these investigation, all of them need not be done during clinical management**.

- To give an example: If chest X-ray and sputum AFB are positive, one should start ATT that need not do costly investigations such as CT scan or even diagnostic laparoscopy, etc. Investigations done by the clinician should be complementary to each other.
  1. **Complete blood picture (CBP)** which include Hb%, TC, DC and ESR. Haemoglobin may be low indicating anaemia. Anaemia has to be corrected before surgery.
  2. **ESR** will help in equivocal cases. High values and the clinical situation may force the clinician to start anti-tuberculous treatment in selected patients. However, with treatment, if ESR comes down and patient is symptomatically better with weight gain, settling fever, improving appetite, it suggests tubercular pathology.
  3. **Sputum AFB (acid fast bacilli):** Demonstrated by Zeihl-Neelsen method. Many patients with abdominal tuberculosis will not have pulmonary tuberculosis. However, if sputum is present, it must be tested for AFB
  4. **Chest X-ray** may suggest tuberculosis in the form of cavity, calcification, etc.

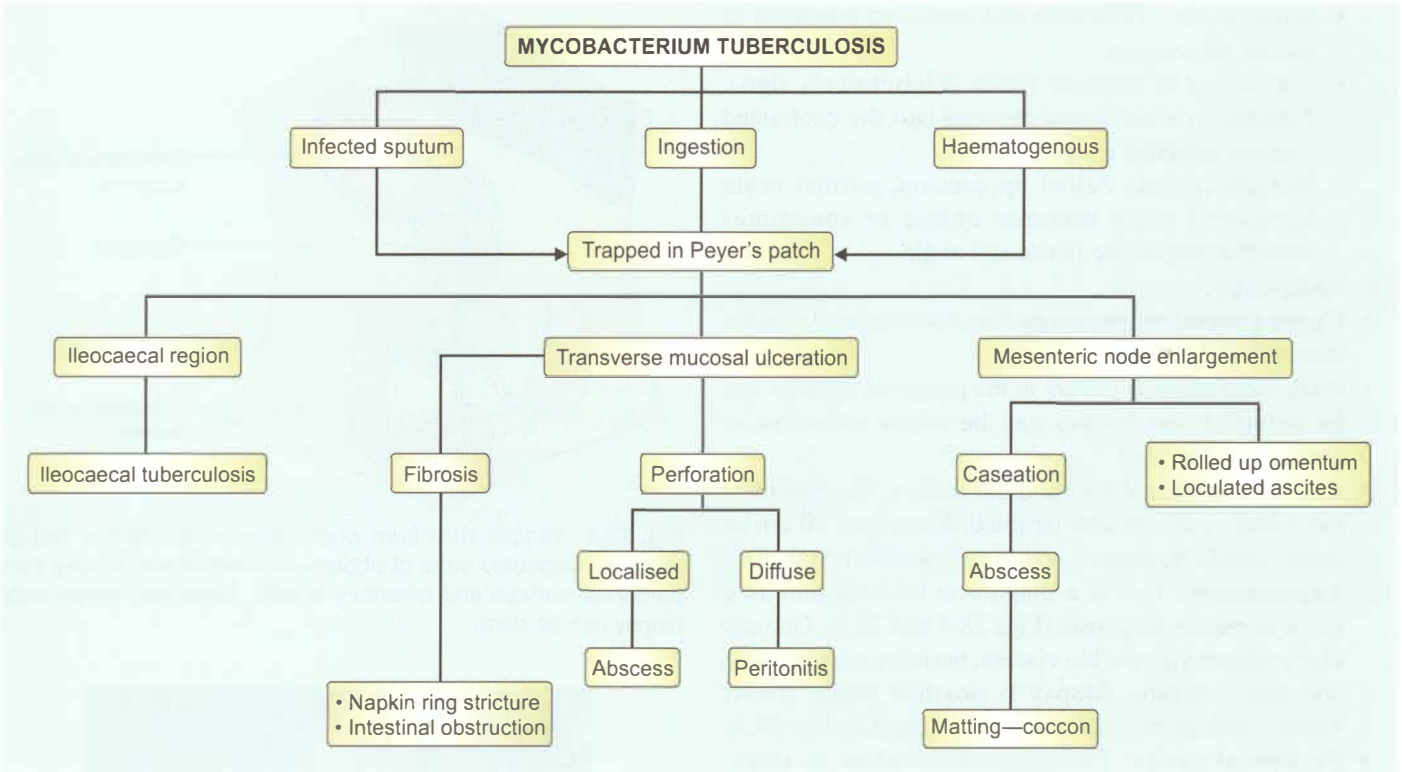


Fig. 28.3: Abdominal tuberculosis—pathology and pathogenesis

5. **Mantoux** test is nonspecific but a strong ulcerated Mantoux test result suggests tuberculosis.
6. **Ultrasound**, being a noninvasive investigation is an imaging of choice.
   - Ascites can be demonstrated and the aspirated fluid is sent for analysis
   - Focal ascites between loops of bowel—**Club sandwich sign** may be seen
   - Enlargement of mesenteric lymph nodes (common) and retroperitoneal nodes (uncommon) can be detected
   - Dilated loops and sometimes peritoneal tubercles are seen as echo-poor shadows
   - Thickening of omentum, mesentery, peritoneum can be found out. (However ultrasound is not the best investigation to detect these findings).
   - **Pseudokidney sign:** Pulled caecum identified in the right hypochondrium
   - Hepatosplenomegaly may be present
7. **Ascitic fluid analysis:**
   - Ultrasound guided fluid is aspirated and about 20–40 ml is sent for analysis (Key Box 28.4).
8. **CECT: Contrast enhanced CT scan of the abdomen:**
   - CT scan is objective. All the findings which can be detected by ultrasound can be confirmed by CT scan. Addition of the contrast is definitely more superior in detecting strictures, dilatations, perforations, and more importantly loculated ascites and intra-abdominal collections. CT guided biopsy can be done. If distension or matting of loops and adhesions are present, it is not safe.
9. **Barium studies:** These are not done routinely. If diagnosis is possible by the various investigations mentioned above, there is no necessity to do them. In fact it can harm the patient by precipitating obstruction and barium peritonitis if there is a perforation.

KEY BOX 28.4

ASCITIC FLUID ANALYSIS (STRAW-COLOURED FLUID)

- Specific gravity is increased—2020 or more
- Glucose < 30 mg%
- AFB is rarely demonstrated <3% of cases
- Increased white cell count (> 500/cells/cumm), predominantly lymphocytic
- Increased total protein (> 2.5 g/dl)
- Serum/ascitic fluid albumin gradient (SAAG*) < 1.1 g% units
- LDH > 90 units/L
- Decreased pH
- Increased adenosine deaminase
- Bacterial isolation and culture is possible in 20–45% of patients.

* **SAAG** = Serum albumin level—ascitic fluid albumin level

- Enteroclysis: Dilatation and narrowed segments in partial obstruction.
- Narrowing of terminal ileum (**Fleischner's sign**), Fibrotic terminal ileum opening into the contracted caecum (**Stierlin's sign**)
- Barium enema: Pulled up caecum, normal acute ileocaecal angle becomes obtuse or sometimes straightening of the ileocaecal angle.

10. **Endoscopy:**
    - **Upper gastroduodenoscopy** may detect tubercles in the stomach or duodenum – rare
    - **Push enteroscopy:** Ulcers in the proximal jejunum can be detected and biopsy can be taken—chances of perforation are high.
    - **Colonoscopy** can detect nodular lesions, ulcerations in the colon—caecum and terminal ileum (last 10 cm of ileum should be entered and biopsy should be taken).
11. **Laparoscopy:** This is a diagnostic investigation as it gives the tissue diagnosis (Figs 28.4 and 28.5). One can also evaluate all possible viscera, peritoneum, omentum and pelvic organs. **Biopsy is possible under direct vision**. Findings may include the following (Key Box 28.5).
    - **Peritoneal cavity:** Fluid collection—clear or straw-coloured or abscess secondary to perforation
    - **Omentum:** Tubercles, rolled up omentum
    - **Peritoneum:** Tubercles, nature of the tubercles, caseation.
    - **Intestines:** Matting of the loops of bowel, adhesions, bands, strictures, dilatations, perforations of bowel
    - **Root of the mesentery:** Short mesentery, caseation of lymph nodes–cold abscess
    - **Lymph nodes:** Mesenteric lymph nodes enlargement, caseation
    - **Caecum:** Pulled up caecum
    - Hepatosplenomegaly, tubercles on the surface of the liver.

    Laparoscopy can also be therapeutic, if a stricture is identified, the diseased loop is isolated, brought out and resection and anastomosis/stricturoplasty done.

KEY BOX 28.5

LAPAROSCOPY—IMPORTANT FINDINGS

- **L**ymph nodes
- **A**dhesions, matting
- **P**eritoneal nodules
- **A**scites – aspiration – AFB staining
- **R**olled up omentum
- **O**bstruction, stricture

You can remember as **LAPARO**

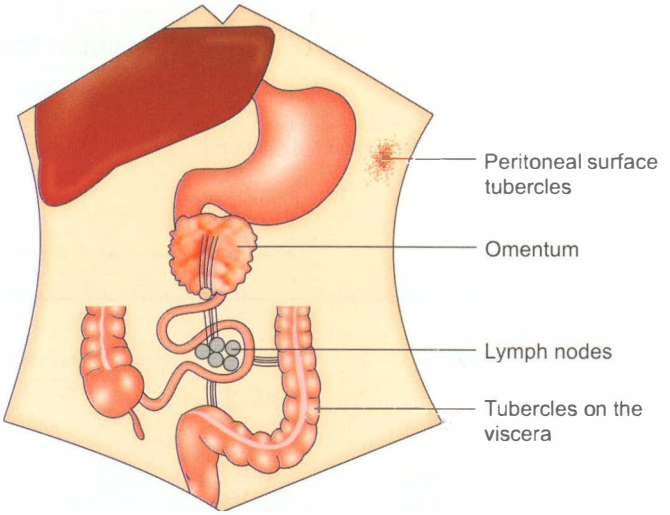


Fig. 28.4: Various sites from where laparoscopic biopsy can be taken in suspected case of abdominal tuberculosis. Biopsy from peritoneal surface and omentum is safe. Otherwise lymph node biopsy can be done

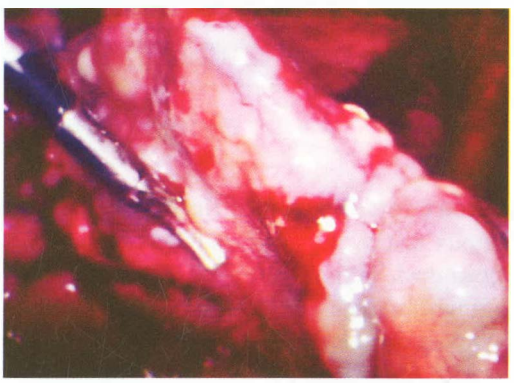


Fig. 28.5: Extensive abdominal tuberculosis. Laparoscopic biopsy being done

12. **Polymerase chain reaction (PCR):**
    - It is a technique used in medical and biological research labs.
    - One can do functional analysis of genes; useful in the diagnosis of hereditary diseases;
    - Helps in detection and diagnosis of infectious diseases such as tuberculosis
    - Laparoscopically biopsied tissue can be sent for PCR. It can detect 1–2 organisms or 8 fg of mycobacterial DNA
    - Positive PCR indicates infection but it need not be active infection. Hence, it is inferior to tissue diagnosis
    - PCR has 97% sensitivity and 99% specificity

When to use what investigations?

In the diagnosis of the abdominal tuberculosis, investigations can be classified as follows.

1. **Routine in all cases:** CBP including ESR, chest X-ray and sputum for AFB. Lymph node biopsy—If lymph nodes are palpable (peripheral—cervical, axillary preferable –

inguinal last choice). Advantage being lymph node biopsy can be done under local anaesthesia. If it is positive, all other invasive and costly investigations can be avoided (*see* clinical notes below). Imaging: Ultrasound examination followed by guided ascitic fluid analysis has become a routine imaging test. It may not be diagnostic but since it has more advantages than any side-effects, it can be done.

2. **Selected cases:** PCR, CT scan, small bowel enteroscopy, barium studies, colonoscopy. If any one of these gives a diagnosis, treatment is started. In the presence of obstruction, avoid barium studies, enteroscopy and colonoscopy. The decision is made on clinical grounds to proceed with laparotomy.
3. **Finally:** Diagnostic laparoscopy/tissue diagnosis—this is the investigation of choice—often done as a last choice when all investigations are equivocal or have failed to give a diagnosis.

Antituberculous Treatment

- Details are given in medicine textbooks. However, 4 drug regimen for 2 months followed by 2 drug regimen for 4 months is recommended as a first line of treatment.
- First line of drugs include INH, rifampicin, ethambutol and pyrazinamide given for 2 months. This is followed by rifampicin and INH for 4 months. Refractory cases are treated by kanamycin, ofloxacin, ciprofloxacin, amikacin, etc. *see* clinical notes.

TUBERCULOUS PERITONITIS

It can be of two types: Acute and chronic. Basically, it produces the following pathological changes:

1. Intense exudation which causes ascitic form
2. Exudation with minimal fibroblastic reaction—loculated form
3. Extensive fibroblastic reaction—plastic form
4. Fibroblastic with secondary infection—purulent form
   - In most of the cases, tuberculous peritonitis results from reactivation of latent primary peritoneal focus.

Types

1. **Ascitic form** (Fig. 28.6) (generalised variety)
   - It is common in children and young adults. The child is brought to the hospital with abdominal distension.
   - Omentum can be felt as a rolled up transverse mass, which is nodular due to extensive fibrosis. Abdomen has a doughy feel with fluid giving rise to shifting dullness.
   - Aspiration of peritoneal fluid reveals exudate, which is rich in lymphocytes (Key Box 28.6).
   - Peritoneal cavity contains pale-straw-coloured fluid and the peritoneal surface is studded with tubercles.
   - Umbilical hernia or congenital hydrocoele appears in children due to increased intra-abdominal pressure.
2. **Loculated or encysted form** (Fig. 28.7)
   - In this variety, ascitic fluid is present in one quadrant of the abdomen which is sealed off by matted intestinal coils surrounded by omentum. It gives rise to localised swelling. These patients have no shifting dullness.
   - It commonly presents in adults.
   - **Differential diagnosis:** Other cystic swellings in the abdomen such as pseudocyst of the pancreas, mesenteric cyst, retroperitoneal cyst.

CLINICAL NOTES

An 18-year-old girl presented with colicky abdominal pain, low grade fever, gross emaciation, etc. Barium meal follow through demonstrated strictures of the small bowel. Laparotomy was done. There were dense adhesions. Minimal dissection resulted in two openings in the terminal ileum. Peritoneum was studded with tubercles from which biopsy was taken. Abdomen was closed with a drainage tube. Postoperatively, the patient was given antituberculous treatment. After three months, the patient came to the outpatient department and had put on 14 kg weight. She was asymptomatic. This case history illustrates the presence of tuberculosis as a major cause of illness in India. It also highlights the importance of antituberculous treatment and recovery.

KEY BOX 28.6

CAUSES OF EXUDATIVE ASCITES—INCREASE IN PROTEIN

Pancreatic ascites
Rare cause: Meig's syndrome
Occlusion: Budd Chiari syndrome
Tubercular peritonitis
Excess of chylomicrons
Infective: Peritonitis
Neoplasm of peritoneum (carcinoma peritonei)
Remember as **PROTEIN**

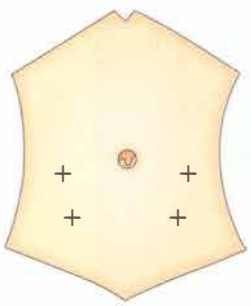


Fig. 28.6: Ascitic form

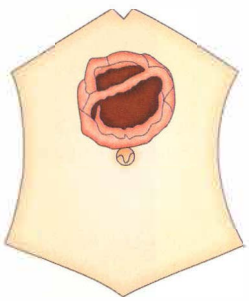


Fig. 28.7: Encysted ascites

3. Fibrous peritonitis (plastic) (Fig. 28.8)

- In this variety, **there is no ascites** but there is extensive fibrosis which results in **dense adhesions** between the coils of intestines. Intestines are matted, distended and not able to empty properly due to adhesions and bands. It is associated with **strictures**.
- This gives rise to blind loop with **steatorrhoea** and **emaciation**.
- Usually, it presents with **intestinal obstruction** at a later date due to fibrous band which needs to be divided to relieve the obstruction. In some occasions, it is not possible to enter the peritoneal cavity, due to dense adhesions.
- It is **not uncommon** to create openings in the bowel at laparotomy and end with a helpless situation wherein one will not be able to close the perforation. The net result is fistula formation.

4. Purulent variety (Fig. 28.9)

- Seen in females as a complication of genitourinary tuberculosis (tuberculous salpingitis).
- The spread occurs through the female genital tract and there is always secondary infection.
- It presents with acute peritonitis at laparotomy, the peritoneal cavity is seen studded with tubercles, cold abscesses and pus.
- Laparotomy, drainage of pus, followed by antituberculous treatment is the choice of therapy.
- It carries poor prognosis because of complications such as toxaemia and faecal fistula formation.
- Tuberculous peritonitis can be associated with infections of pleural space and pericardial space (effusion). It is called **polyserositis** syndrome.
- *See* also Figs 28.10 to 28.13

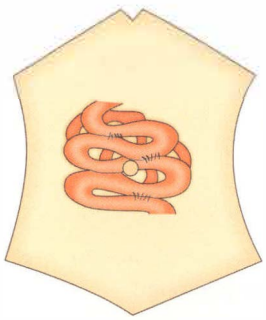


Fig. 28.8: Fibrous peritonitis

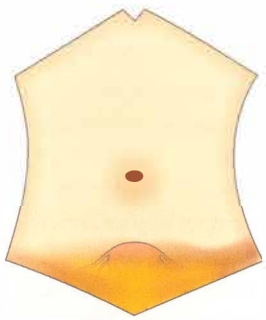


Fig. 28.9: Purulent type

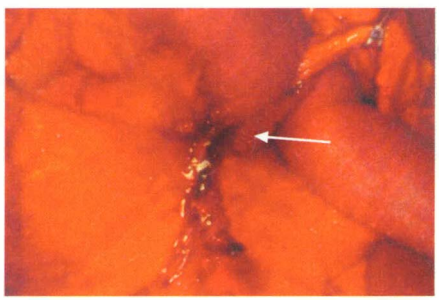


Fig. 28.10: Fibrous bands

Fig. 28.11: Adhesive form

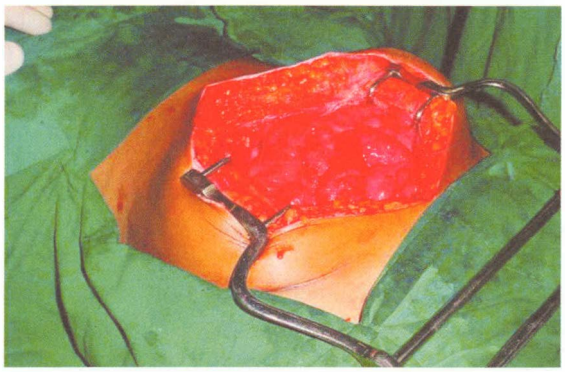


Fig. 28.12: Tubercles on the peritoneal surface

Fig. 28.13: Tuberculous peritonitis

TUBERCULOUS MESENTERIC LYMPHADENITIS

Clinical presentation

1. As a calcified lesion (Fig. 28.14) along the line of mesentery, which extends from L2 vertebra, at the left of vertebral column to the right sacroiliac joint. In 50% of cases, there is no active infection but in the remaining, there is infection. If the symptoms are that of tuberculosis, antituberculous treatment should be given. The shadow caused by lymph nodes are round to oval, mottled and may be regular or irregular.

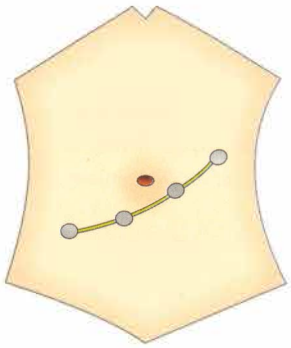


Fig. 28.14: Mesenteric lymph nodes

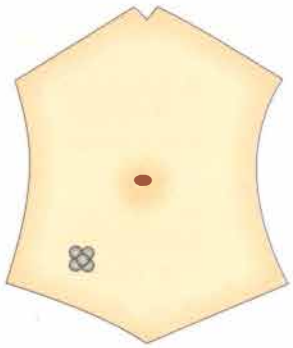


Fig. 28.15: Acute lymphadenitis

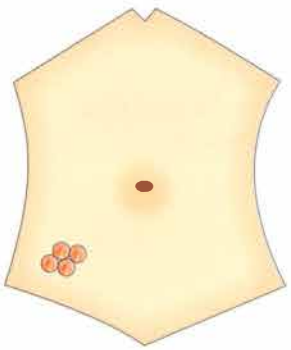


Fig. 28.16: Chronic lymphadenitis

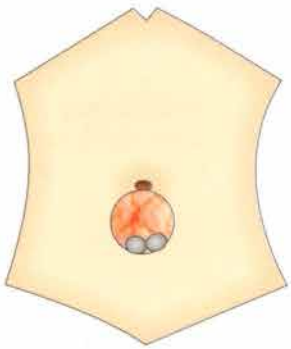


Fig. 28.17: Pseudomesenteric cyst

2. Acute mesenteric lymphadenitis (Fig. 28.15)

- Common in children, clinically mimics acute appendicitis.
- Pain in the right iliac fossa, vomiting, fever, rigidity can be present.
- On palpation, tender mass of swollen lymph nodes can be felt in the right iliac fossa.
- Laparotomy, appendicectomy and biopsy of the lymph node is the procedure of choice.

3. Chronic lymphadenitis (Fig. 28.16) in children presents as **failure to thrive**. Fever, loss of weight, loss of appetite, emaciation and pallor are present. Abdomen is protuberant. On deep palpation, nodes can be felt in the right iliac fossa. These nodes have to be differentiated from nodes that enlarge due to lymphoma.

4. Pseudomesenteric cyst (Fig. 28.17)

- This is due to caseation of mesenteric lymph nodes confined within two leaves of mesentery.
- Due to adhesions, intestines can get kinked or twisted causing intestinal obstruction.

Treatment

- Antituberculous treatment (details in medicine books).

INTESTINAL TUBERCULOSIS

Ileocaecal region is commonly involved in tuberculosis because of the following reasons:

1. Rich lymphatics in Peyer's patches.
2. Presence of alkaline media favours the growth of the organisms.
3. Presence of ileocaecal valve precipitates stasis.
4. Terminal ileum is the area of maximum absorption.

Types (Table 28.1 for comparison)

1. Ulcerative variety
2. Hyperplastic variety
3. Mixed (sometimes)

Clinical features (Figs 28.18 to 28.20)

In general, intestinal tuberculosis presents with the following symptoms:

- **Abdominal pain:** It is the most common symptom. It can be a dull, vague pain or colicky pain (stricture) which increases after taking food or relieved by vomiting.
- **Diarrhoea:** Watery, small quantity, abnormally foul smelling. It may alternate with constipation.
- **Nonspecific symptoms** such as flatulence, noisy sounds in the abdomen (**borborygmi**) are not uncommon.
- **Abdominal distension:** It is due to ascites and subacute intestinal obstruction.
- Weight loss is very common. Anorexia, tiredness, pallor may be the presenting features.

Signs

- Typically patients are **malnourished and pale**.
- **Visible intestinal peristalsis** may be seen.
- **Distended bowel loops** can be palpated.
- **Doughy abdomen** in case of peritoneal involvement.
- **Rolled up omentum,** mass in the right iliac fossa or lumbar region, loculated ascites, etc. are other features (Fig. 28.21).

Management of intestinal tuberculosis

1. **No evidence of intestinal obstruction:** Antituberculous treatment.
2. **With obstruction (stricture)** (Figs 28.22 and 28.23)
   - **A. Solitary stricture:** It is best treated by stricturoplasty by incising the stricture longitudinally and suturing it transversely (Fig. 28.28).
   - **B. Multiple strictures** at long intervals: Stricturoplasty is the ideal treatment.
   - **C. Multiple strictures within a short segment.** Resection is the ideal treatment (Fig. 28.30).

PEARLS OF WISDOM

Stricture within 10 cm from the ileocaecal junction is best resected.

Table 28.1 Comparison of two forms of intestinal tuberculosis (TB)

	Ulcerative variety	Hyperplastic variety
1. Aetiology	Secondary to pulmonary TB. Occurs due to swallowing of TB bacilli (*Mycobacterium TB*)	It is a primary intestinal TB, due to *M. bovis*. Milk-borne infection or due to *Mycobacterium TB*—low grade infection
2. Site	Terminal ileum	Ileocaecal region
3. Virulence of the organism	More virulent	Less virulent
4. Resistance of body	**Very poor**	Good
5. Pathology	Multiple ulcerations in the terminal ileum with/without involvement of lymph nodes. **Ulcers are transverse.** Serosa is reddened and oedematous.	It is **low grade, chronic continuous inflammation** involving ileocaecal region resulting in cicatrising **granuloma** in right iliac fossa (mass in right iliac fossa).
6. Clinical features	Symptoms of **pulmonary TB,** blood and mucus in stool resulting in gross emaciation and cachexia. Diarrhoea is also a feature.	Abdominal pain and diarrhoea may be the initial symptoms for a long time and later fever, weight loss and subacute intestinal obstruction occur.
7. Complications	• **Acute** TB, ulcer perforation—ulcers are transverse because they follow the lymphatics. Treatment is laparotomy and resection of bowel. • **Chronic:** Healing of ulcer results in **stricture** of terminal ileum and **subacute intestinal obstruction**.	Nodular, mobile, **firm mass** in the right iliac fossa which later produces subacute **intestinal obstruction**.
8. Barium meal follow through (Figs 28.24 and 28.25)	Demonstrates a stricture, or multiple strictures. In the initial stages, ileum is not seen due to hypermotility.	**Barium enema** can demonstrate (i) contracted caecum, (ii) pulled up caecum (subhepatic), (iii) luminal obstruction, and (iv) obtuse ileocaecal angle.
9. Chest X-ray and sputum AFB (Fig. 28.26)	Positive	Negative
10. **Colonoscopy and biopsy** (Fig. 28.27)	To confirm the diagnosis	To confirm the diagnosis

Fig. 28.18: Step ladder peristalsis is seen in a patient who underwent appendectomy for pain in the right iliac fossa. The diagnosis made for acute appendicitis and appendicectomy was done. Postoperatively patient has step ladder peristalsis. The case was ileocaecal tuberculosis

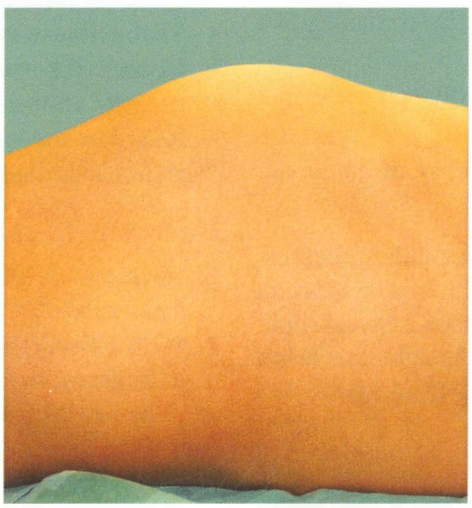


Fig. 28.19: Distension of the abdomen due to ascites

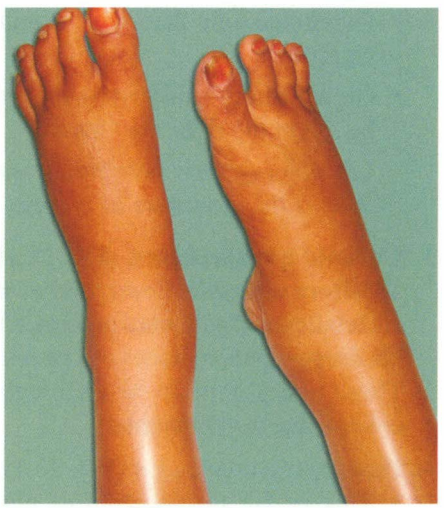


Fig. 28.20: Severe hypoproteinaemia—you can see bilateral pedal oedema

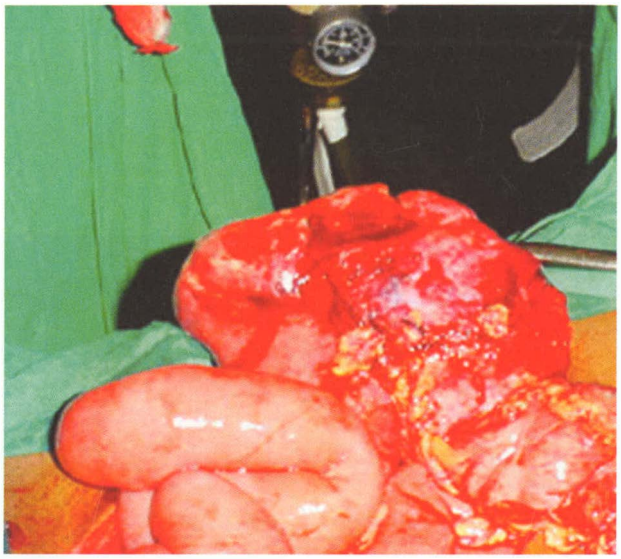
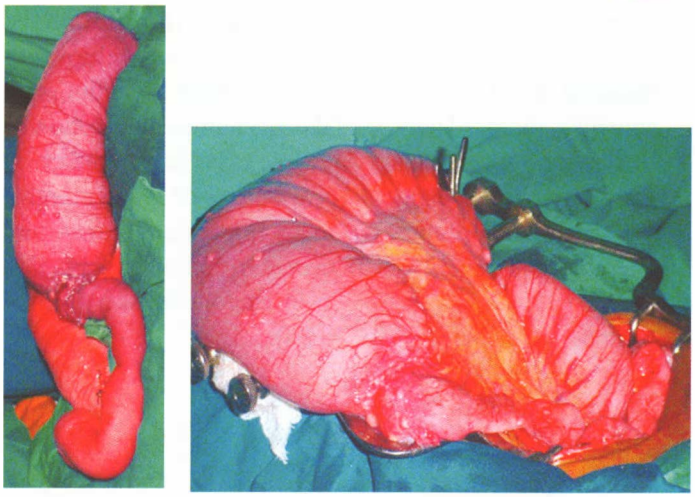


Fig. 28.21: Mass in the right iliac fossa—hyperplastic variety



Figs 28.22 and 28.23: Tuberculous stricture with intestinal obstruction—observe the stricture, tubercles and massive proximal dilatation

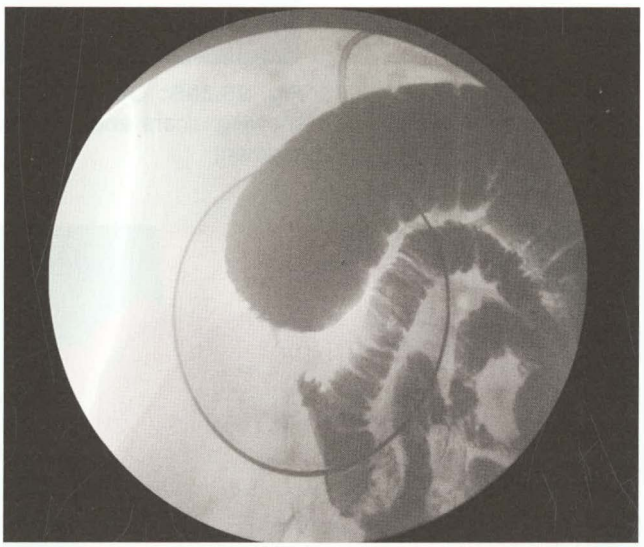


Fig. 28.24: Enteroclysis showing stricture of the jejunum with proximal dilatation

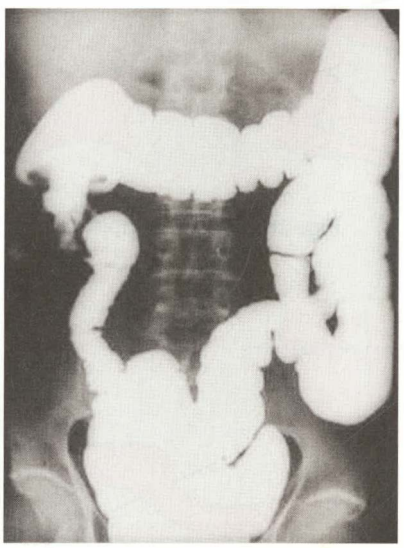


Fig. 28.25: Barium enema—'pulled up' caecum

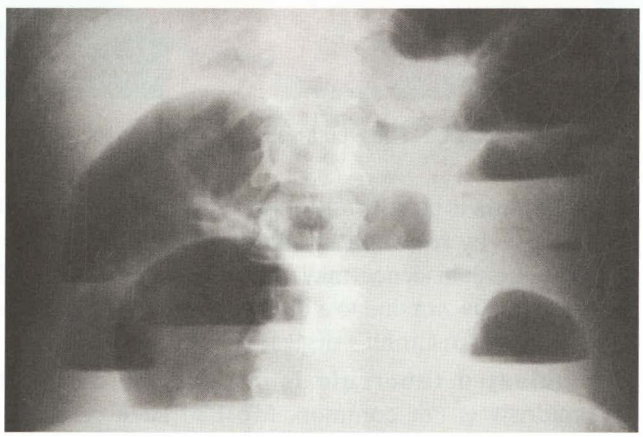


Fig. 28.26: Plain X-ray showing dilated air fluid levels

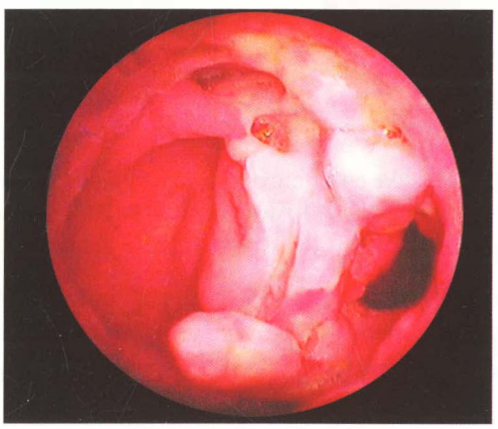


Fig. 28.27: Colonoscopy showing nodular leison in the ascending colon. Biopsy proved tuberculosis

3. **Surgical treatment of hyperplastic tuberculosis:** Limited resection is the treatment of choice. It includes removal of terminal 8–10 cm of the diseased ileum, caecum with appendix, diseased portion of the ascending colon, followed by ileocolic anastomosis. All these cases have to be given antituberculous treatment for a period of 9–12 months. Nutritional supplementation to improve albumin and haemoglobin levels and if necessary, blood transfusion before and after surgery help in smooth recovery in the postoperative period (Fig. 28.29).

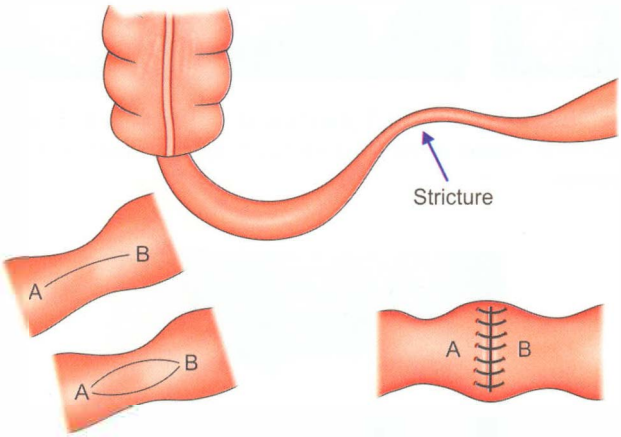


Fig. 28.28: Stricturoplasty

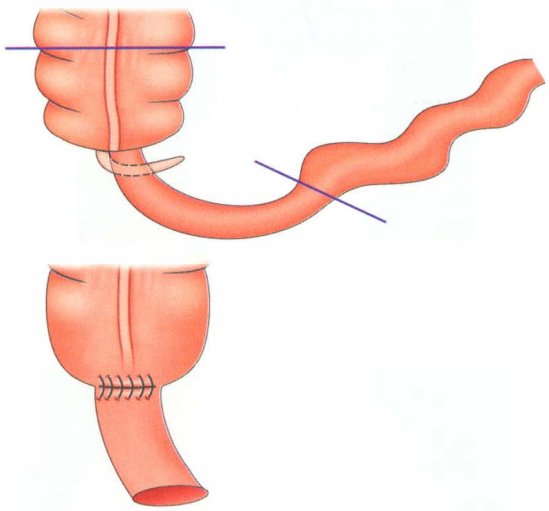


Fig. 28.29: Limited colectomy followed by ileocolic anastomosis. (Courtesy: Miss Vidushi Kapil, MBBS student, KMC, Manipal)

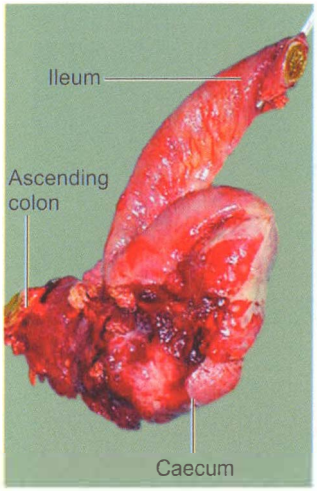


Fig. 28.30A: Limited resection—resection of the diseased segment

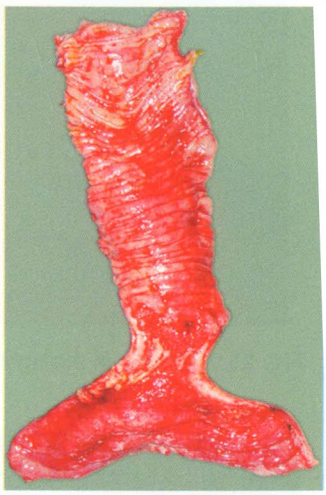


Fig. 28.30B: Opened bowel showing ulcers and narrowed segment

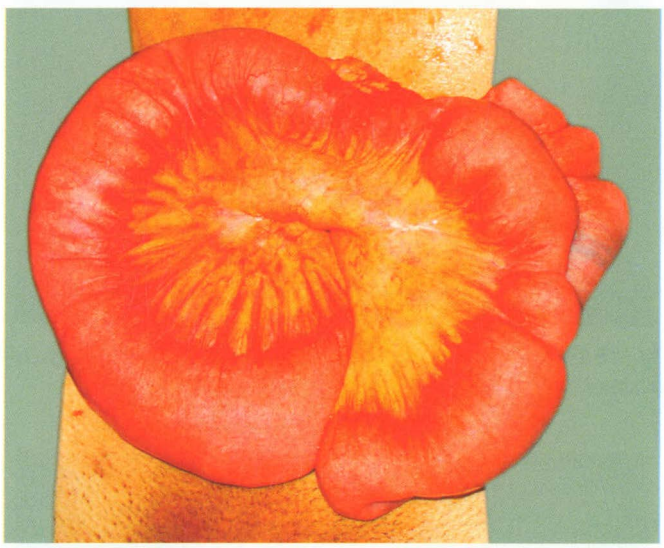


Fig. 28.30C: Tubercular stricture of jejunum

Complications of abdominal tuberculosis

1. **Intestinal obstruction:** Usually it is ileal or jejunal obstruction—details have been already discussed
2. **Perforation:** It is not uncommon. Carries 6–8% mortality rate specially in late cases with peritonitis.
3. **Malnutrition:** Diarrhoea, loose stools, blind loop syndromes contribute for malnutrition
4. **Faecal fistula** is usually due to operated cases of intestinal tuberculosis by resection and anastomosis. In such cases, anastomotic dehiscence may result in faecal fistula. The faecal fistula is not due to a direct internal involvement of viscera unlike in Crohn's disease.
5. **Disseminated tuberculosis** *per se* from abdominal tuberculosis is not common. Most of such cases have fulminant pulmonary tuberculosis. This happens in untreated cases and immunocompromised cases.

INFLAMMATORY BOWEL DISEASES

Introduction

These are the diseases involving small and large bowel, of unknown aetiology, characterised by multiple ulcerations in the bowel, clinically manifesting as blood and mucus in stools. Ulcerative colitis and Crohn's disease (regional enteritis) are included under this heading. However, both these diseases are uncommon in India. Hence, amoebic dysentery, intestinal TB, enteric fever with intestinal manifestations can also be included in this. Both diseases can present has acute abdomen with intestinal obstruction and complications such as perforation toxaemia, etc. Also both are premalignant conditions for carcinoma colon.

ULCERATIVE COLITIS

Aetiology

1. **Autoimmune factor:** Even though exact mechanism of ulcerative colitis is not clear, there are some factors which may point out to autoimmune reaction. They are:
   - Presence of **cytotoxic T lymphocytes** against colonic epithelial cells in the lamina propria of the bowel.
   - Presence of **anticolon antibodies**.
   - Whatever be the immune mechanisms, activation of inflammatory mediators such as cytokines, growth factors and arachidonic acids takes place and they are responsible for the disease.
2. **Dysfunctional immunoregulation** in the intestinal wall results in inappropriate production of **cytokines**. This creates an imbalance between various interleukins resulting in inflammatory changes.
3. **Psychosomatic and personality factors:** Ulcerative colitis is more common in western women. Emotional stress, family stress, stress from divorce are the contributing factors.

PEARLS OF WISDOM

Western, white, worrying women's disease is ulcerative colitis.

4. **Dietary factors**
   - Westernisation of the diet which is rich in red meat has been blamed. Vegetarian diet is supposed to protect the colonic mucosa.
   - **Allergy to milk protein** is responsible for ulcerative colitis in a few patients.
5. **Defective mucin production** and a defective mucosal immunological reaction is considered as a chief factor responsible for ulcerative colitis.

PEARLS OF WISDOM

Appendicectomy and smoking have been protective factors for development of ulcerative colitis.

6. **Genetic:** 15% of patients have first degree relatives with ulcerative colitis.

Pathology (Key Box 28.7)

- The disease always starts in the rectum and spreads in a backward manner, thus involving the entire colon in majority of cases. In 5% of cases, terminal ileum can also be involved—**back wash ileitis**.
- **Anus is not involved in ulcerative colitis.**
- The disease manifests as multiple, small superficial ulcers—**pinpoint ulcers**.
- As the disease progresses, inflammation spreads into the submucosa of the colon. Attempt at healing may produce **pseudopolyp**. There are areas of epithelial hypertrophy in between the ulcers, resembling polyp. Healing with fibrosis results in a narrow, contracted colon, called **pipe stem colon**.
- Microscopy: **Pus** (abscess) in the crypts and pus cells (inflammatory cells) in the lamina propria are typical of ulcerative colitis.

Clinical features

- More common in females. Female:male ratio is 2:1.
- Age: Common age of presentation is 3rd decade followed by 4th and 2nd decades.
- The disease is characterised by passage of 15–20 stools per day and contains blood and mucus. Sometimes, it may be watery diarrhoea (Key Box 28.8). As the rectum loses elasticity and lumen collapses, tenesmus occurs.
- Relapses and remissions are common and are related to emotional disturbances.

KEY BOX 28.7

PATHOLOGY OF ULCERATIVE COLITIS

Pinpoint ulcers
Pseudopolyposis
Pipe stem colon
Pus cells

Observe **4 Ps**

KEY BOX 28.8

TYPES—DEPENDING UPON SEVERITY

- **Mild colitis:** Refers to < 4 stools/day without systemic signs and symptoms.
- **Moderate colitis:** Refers to < 6 stools/day without systemic toxicity.
- **Severe colitis:** Refers to > 6 stools/day with systemic toxicity—fever over 37.5°C, tachycardia more than 90/min, hypoalbuminaemia less than 3 g/dl, weight loss more than 3 kg

- Severe dehydration, malnutrition, anaemia, hypoproteinaemia are late features.
- Acute fulminating attack is associated with high grade fever, bloody dysentery, distension and tenderness all over the abdomen with profound weakness. Hypokalaemia, acidosis, anaemia and shock are the other features.

Types of ulcerative colitis (depending upon the extent of the colon involved)

1. **Proctitis:** In about 20–25% of the patients, the disease involves only rectum. In such patients, stools are semisolid because of absorption of water by normal colon. Also, the intensity of the disease is not severe and risk of cancer is 2–5%.
2. **Left-sided colitis:** It is found in 15% of patients. It presents as severe recurrent attacks of diarrhoea with blood in stools, without systemic toxicity (Fig. 28.31A).
3. **Total proctocolitis:** It is seen in about 25% of the patients. Severe bloody diarrhoea and hypoproteinaemia are its features. Chances of cancer and complications are high in this group (Fig. 28.31B).

Complications

1. **Toxic megacolon:** It is an abdominal emergency encountered with fulminating colitis. Severe abdominal pain and tenderness, toxaemia, high fever, tachycardia and leucocytosis are the features. Plain X-ray abdomen which shows colon with a diameter more than 6 cm gives the diagnosis. It requires emergency treatment by laparotomy and colectomy. Supportive treatment and intravenous corticosteroids are necessary (Key Boxes 28.9 and 28.10, Figs 28.32 and 28.33).

Fig. 28.32: Plain X-ray abdomen showing dilated transverse colon more than 6 cm toxic megacolon

KEY BOX 28.9

FACTORS PRECIPITATING TOXIC MEGACOLON

- Barium enema
- Opiates, antidiarrhoeal drugs, anticholinergic agents
- Not known

KEY BOX 28.10

TOXIC MEGACOLON—CAUSES

- Ulcerative colitis, salmonella colitis
- Pseudomembranous colitis, amoebic colitis

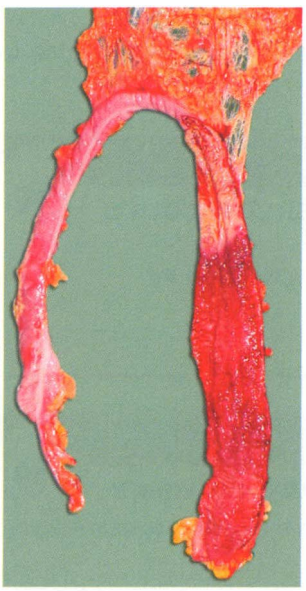


Fig. 28.31A: Sharp demarcation at the splenic flexure may be due to an incomplete marginal artery ending at point of demarcation. Case of left-sided colitis

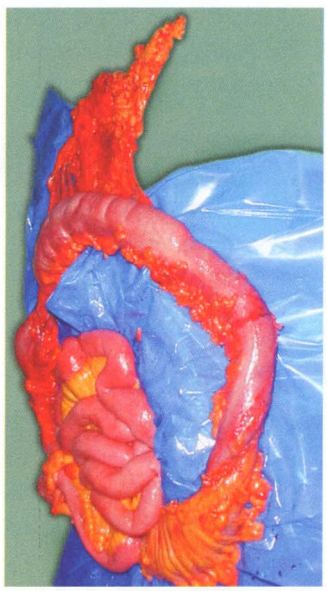


Fig. 28.31B: Fully mobilised colon. Observe loss of haustrations. Total proctocolectomy with ileal pouch to anal anastomosis was done—total proctocolitis

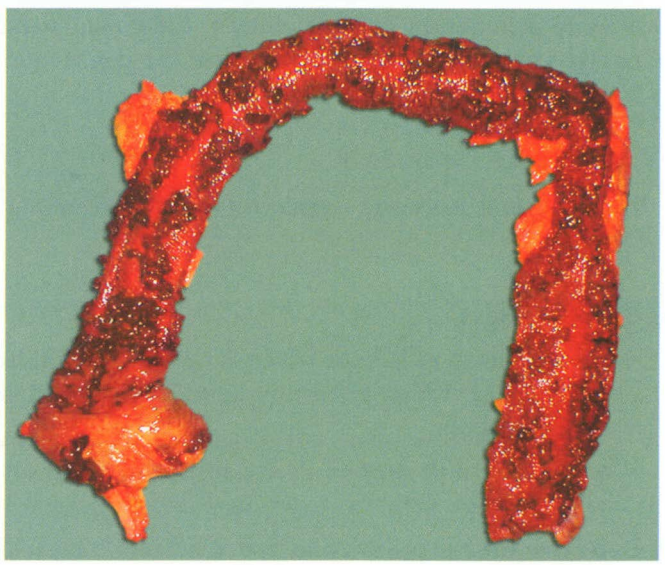


Fig. 28.33: Total colectomy for toxic megacolon specimen showing pseudopolyposis

2. **Massive haemorrhage** per rectum is uncommon. It is treated by blood transfusion.
3. **Perforation** is treated as peritonitis with resection of colon. Mortality rate is around 25–50%. Steroids may mask the symptoms. In an emergency situation, saving life is more important. Hence, resection followed by ileostomy should be done.
4. **Carcinoma of the colon** (Key Box 28.11)
   - Overall incidence is 3% when the disease has been present for 15 years.
   - At the end of 25 years, the incidence may be around 20%.
   - Hence, routine sigmoidoscopy and biopsy have to be done when the disease is present for more than 10 years and if it shows epithelial dysplasia, it should be considered as premalignant.
   - Incidence is more in total proctocolitis and when the disease has started in the early age group.
5. **Recurrent perianal abscess** resulting in perianal fistula, in about 15–20% of patients.
6. **General complications (extraintestinal)**
   - **Protein malnutrition** resulting in cirrhosis. Primary sclerosing cholangitis is also found in many cases. Fatty acid infiltration is seen in 40% of cases. It is reversible after control of disease.
   - **Skin ulcerations**, pyoderma, erythema nodosum, etc. reflects protein malnutrition.
   - Conjunctivitis, iritis, arthritis involving large joints are also other features.
   - Incidence of **bile duct cancer** is high in these patients.

PEARLS OF WISDOM

Peripheral arthritis and ankylosing spondylitis are the two most common extraintestinal manifestations.

Differential diagnosis

- **Crohn's disease** should be ruled out first. The differences between the two inflammatory bowel diseases have been discussed at the end of the chapter. In general, diarrhoea and bleeding are more common with ulcerative colitis than Crohn's disease.
- **Dysenteries:** Bacillary dysenteries, Shigellosis, Salmonellosis, Amoebiasis and other dysenteries have to be kept in mind specially in developing countries including tuberculosis which has been already discussed.
- **Diverticular disease of the colon:** However, when complications including bleeding occur, diverticular disease of the colon and in cases of perforations, malignant perforation must be considered as a differential diagnosis.

Investigations

1. **Stool**
   - It is done mainly to rule out various causes of infective diarrhoea—amoebiasis, Shigella, Clostridium difficile.
   - Most common cause of infective colitis in UK is Campylobacterium.
2. **Sigmoidoscopy** (Key Box 28.12)
   - Can demonstrate inflammatory changes in the mucosa.
   - Mucus, pus and blood are visible.
   - Multiple ulcers are visible with bleeding.
3. **Barium enema findings in ulcerative colitis** (should not be done in acute cases) (Fig. 28.34)
   - Contracted colon/pipe stem colon
   - Absence of haustrations and mucosal irregularity
   - Pseudopolyposis appears as stippled appearance

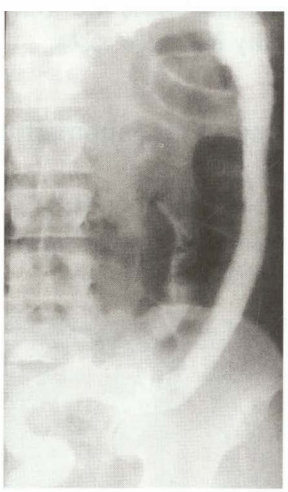


Fig. 28.34: Barium enema showing pipe stem colon

KEY BOX 28.11

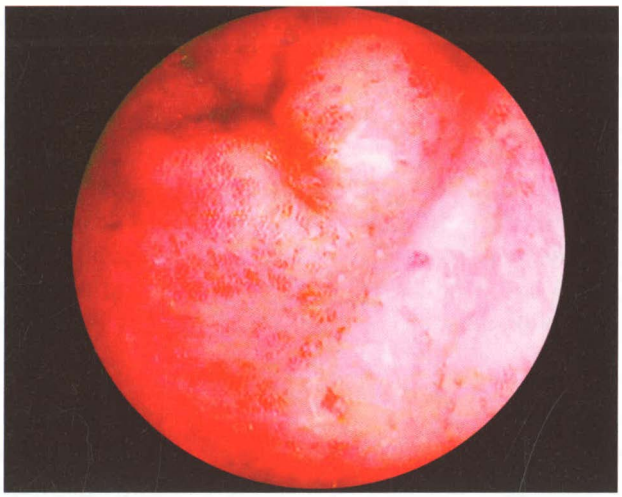
ULCERATIVE COLITIS (UC) AND COLORECTAL CANCER (CRC)

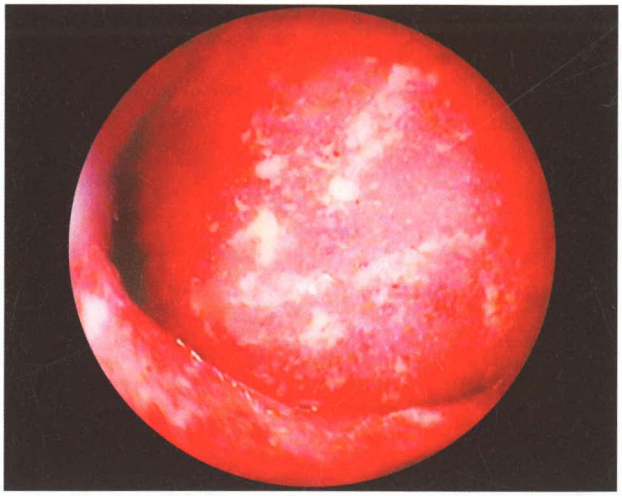
- CRC is more aggressive
- Multicentric and synchronous
- UC patients with primary sclerosing cholangitis (PSC) have increased risk of CRC
- More advanced stage at the time of presentation
- Risk increases with duration of the disease
- Malignancy develops on a background of dysplasia [dysplasia associated lesion or mass **(DAL-M)**]

KEY BOX 28.12

SIGMOIDOSCOPY—FINDINGS

Amoebic ulcer	Ulcerative colitis
• Large	• Small, pinpoint
• Deep, flask shaped	• Superficial
• Mucosa in between ulcer is healthy	• Mucosa in between ulcers unhealthy





Figs 28.35 and 28.36: Colonoscopy—pinpoint ulcers with bleeding (*Courtesy:* Dr Filipe Alvares, Medical Gastroenterologist, KMC, Manipal)

4. **Colonoscopy** (Figs 28.35 and 28.36)
   - To confirm the diagnosis by biopsy
   - To find out the extent of involvement of colon
   - To follow up patients who are on treatment
   - To rule out carcinomatous changes
5. **Plain X-ray abdomen:** To rule out megacolon and perforation.
6. **C-reactive protein:** Its levels are very high in case of acute fulminating attack or toxic megacolon.
7. Electrolytes, albumin levels are low. They have to be corrected especially in severe cases.

Treatment of ulcerative colitis

I. Conservative line of management (Table 28.2)

- Hospitalisation and bedrest
- Correction of fluid and electrolyte imbalance
- Blood transfusions to correct anaemia and TPN for hypoproteinaemia (Fig. 28.37).

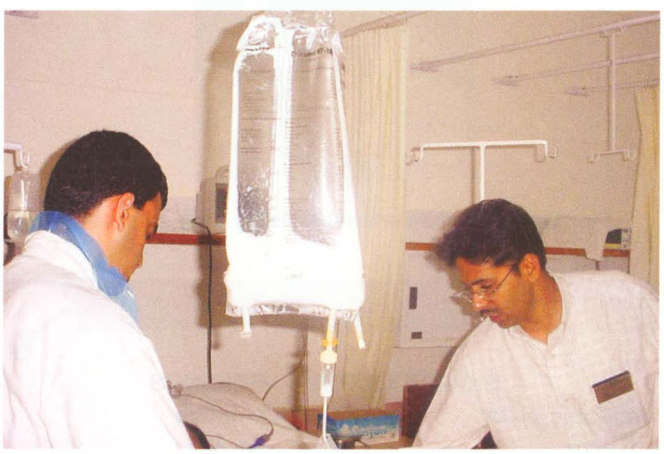


Fig. 28.37: Total parenteral nutrition (TPN) before and after surgery for ulcerative colitis plays an important role in the recovery of the patient, as these patients are grossly emaciated and hypoproteinaemic

- **Salazopyrines** are given in the dose of 2 g/day. Mode of action: When given orally, it gets split into 5-amino-salicylic acid and sulphapyridine in the colon. This suppresses activity of prostaglandins E1 and E2 and thus reduces inflammation. They are used mainly to induce remission.
- **Corticosteroids:** Less severe cases not responding to salazopyrines are given a trial of oral prednisolone 60 mg/day. They decrease the frequency of stools. The dose is tapered off over 3–4 weeks.
- In acute attacks, IV hydrocortisone 100 mg is given.
- Prednisolone retention enema: 20 mg in 200 ml saline in intractable diarrhoea. It avoids systemic toxicity. Prednisolone 20–40 mg/day can also be given orally for 3–4 weeks.
- **Role of cyclosporine:** Those patients who do not respond to corticosteroids, can be given IV cyclosporin 4 mg/kg/day. It can induce remission.

II. Surgery

Indications for surgery

- Complications—toxic megacolon, perforation.
- Active disease in spite of medical line of management
- Severe disease not responding to medical treatment
- Dysplasia on biopsy
- Steroid dependence
- Haemorrhage

PEARLS OF WISDOM

Failure to improve within 7 days of maximum medical treatment with steroids and cyclosporine is an indication for surgery.

Table 28.2 Pharmacotherapy in ulcerative colitis

Drug	Dose	Mode of action
1. Salazopyrines—**first choice**	• 2–4 g/day • Anti-inflammatory • Immunosuppressive	• Suppresses PGE_1 and E_2
2. Steroids—refractory cases	• IV hydrocortisone 100 mg 8th hourly • Prednisolone enema • After acute illness prednisolone 40–60 mg/day	• Anti-inflammatory
3. Immunomodulators • Cyclosporine used in refractory cases	• Azathioprine 6-mercaptopurines	• Act at DNA level and inhibit T-lymphocyte functions
4. Monoclonal antibody – Antitumour necrosis factor-alpha-(TNF α) Infliximab		• Antitumour necrosis
5. Anti-inflammatory (salicylic acid)	• Olsalazine (Dipentum) • 500 mg 1–2 times/day	• Directly targeted/released into the colon

1. **Total proctocolectomy** followed by permanent ileostomy (ileoanal anastomosis should not be done because of incontinence). Ileostomy is connected to ileostomy bag. Adhesive obstruction and chronic perineal sinus are late complications. This is the procedure with least complications (Figs 28.38 and 28.39).
2. **Restorative proctocolectomy** with ileal pouch:

- This can be done as one- or two- stage procedure.
- Total proctocolectomy is done first.
- A mucosectomy of the upper anal canal is done.
- A pouch is created by anastomosing the loops of ileum. A J shaped pouch is the most popular followed by W pouch (Figs 28.40 to 28.43).
- The pouch is anastomosed to the dentate line (junction of upper and lower anal canal) by using stapler or by hand sutures.
- Protective ileostomy is done and it can be closed after two months.

Advantages of a pouch

1. Avoids ileostomy

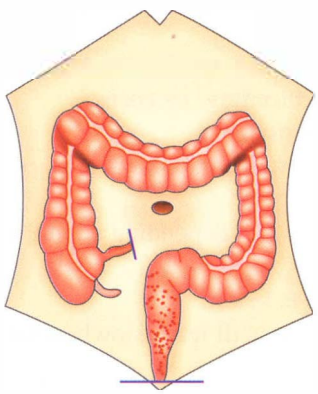


Fig. 28.38: Total colectomy

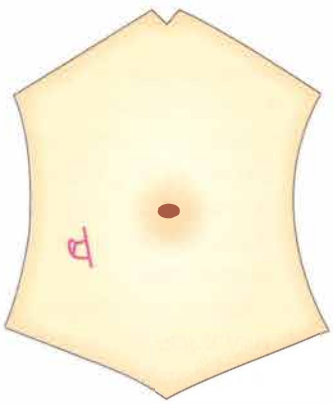


Fig. 28.39: Ileostomy

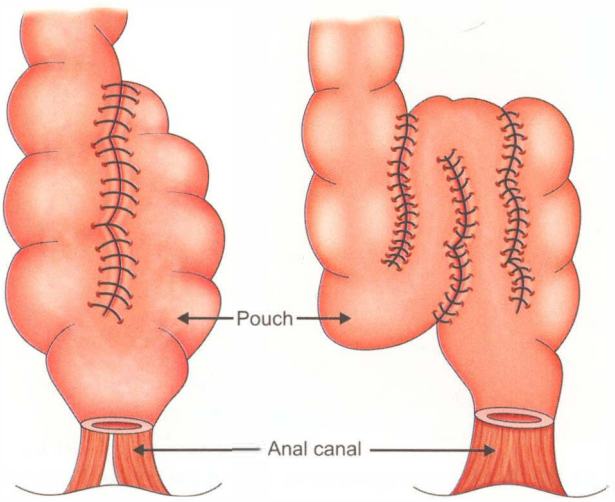


Fig. 28.40: J pouch

Fig. 28.41: W pouch

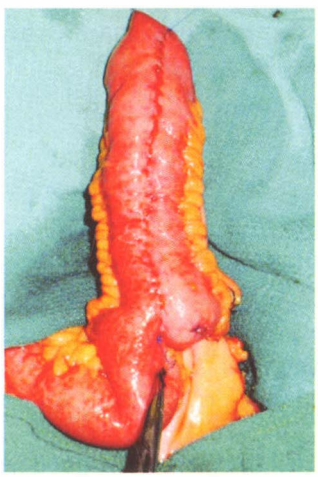


Fig. 28.42: J pouch (*Courtesy:* Dr Prasad Babu TLVD Ramcharan Thyagarajan, Srikanth Gadiyaram, Prof Sadiq S Sikora, Department of Surgical Gastroenterology, Manipal Hospital, Bengaluru)

The length of the pouch is generally 15 cm and capacity around 300 ml when distended. Note that it is a two-layered anastomosis to create the pouch. The same can be achieved by linear stapling device.

Table 28.3 Treatment of ulcerative colitis

	Mild	Moderate	Severe
Admission	Not required	Not required	Required
Rectal steroid	Yes	Yes	No
Prednisolone	Oral 20–40 mg	Yes	IV hydrocortisone 100–200 mg/4 times/day
5 ASA	Can be given	Can be given	Not given
Nutrition	**Oral**	**Oral**	**TPN/oral/high calorie diet**
Fluid/electrolyte balance	Normal	Normal	Imbalance is very common
Blood transfusion	No	Yes	Yes
Surgery	Not done	Not done	No response for 3–5 days—**surgery**

5ASA: **amino salicylic acid**
TPN: **total parenteral nutrition**

Wound was sutured after 48 hours because wound infection is a common complication after this surgery

Fig. 28.43: Total proctocolectomy with a pouch and protective ileostomy was done here

2. Continence is preserved and patient is able to pass the stools *via* naturalis.
3. At the same time, all the diseased **mucosa** has been removed. Thus, the risk of cancer is negligible.
   - Pouchitis is a complication.
   - *See* Table 28.3 also.

Prognosis

- In general, emergency colitis is required in about 25% patients with severe attacks. Perforation has a mortality rate of up to 40% because of faecal contents and toxicity. Incidence of carcinoma colon is about 10–15%. Longer the duration of the disease, higher is the incidence.

Dietary advice

- High protein, carbohydrates, whole grains, and good fats. Meat, fish, poultry, and dairy products breads and cereals; fruits and vegetables may be consumed.
- For vegetarians: Dairy products and plant proteins — such as soya bean products may be consumed.
- To avoid: High fibre high residue diet –thus to control diarrhoea
- To avoid: Caffeine-Coffee, dried fruits and nuts, alcohol, meat, spicy food, oily food, soda, etc.

ILEOSTOMY

Definition

- Ileostomy is a surgical procedure wherein a loop of the ileum or end of the ileum is brought to the exterior (surface of the body).
- Two types are usually done: End ileostomy and loop ileosotmy.
- End ileostomy is done following total proctocolectomy. It is a permanent ileostomy. The ileum is brought out through the lateral edge of rectus abdominis. It should project at least 5 cm outside (Fig. 28.44).
- Loop ileostomy is done to divert gastrointestinal contents to protect ileo-pouch anastomosis. It is a temporary ileostomy which is closed after 6–8 weeks.
- Permanent ileostomy is also required following total colectomy for carcinoma colon. It is an end ileostomy (Figs 28.45 to 28.47).
- Ileostomy care includes maintenance of fluid and electrolyte balance, use of disposable ileostomy bag and skin protection.
- Complications of an ileostomy are similar to that of colostomy-retraction, prolapse, bleeding, stenosis.
- Precautions—living with an ileostomy
  1. Ileostomy bag has to be fitted well to the body surface. It needs to be adjusted often
  2. Ileostomy has to emptied 4–6 times depending upon the requirement
  3. Chewed and masticated food is better
  4. Avoid gas forming diet. Patients will learn slowly what to take and what not to take
  5. Long-term complications include—gall stones, kidney stones, adhesions and intestinal obstruction.

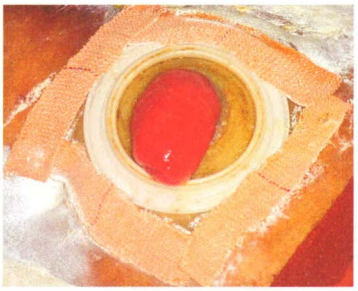


Fig. 28.44A: Ileostomy: It should project out for at least 5 cm

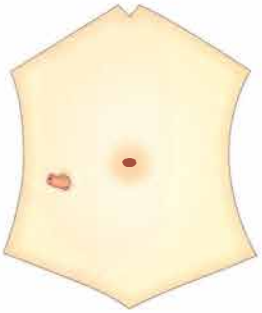


Fig. 28.44B: End ileostomy: following total protocolectomy

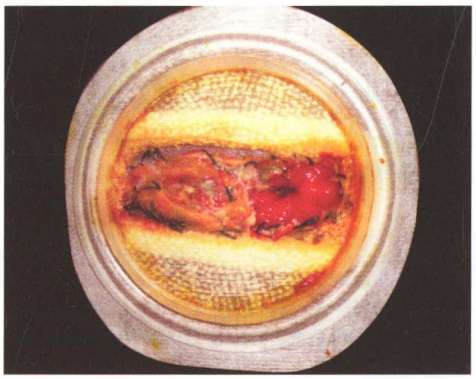


Fig. 28.45: Doubtful viability—complication of the ileostomy

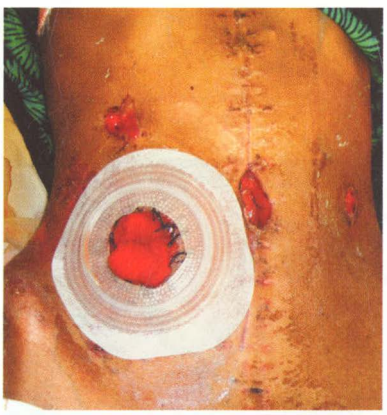


Fig. 28.46: 18-year-old girl was diagnosed with acute appendicitis at exploration, tuberculosis of the ileum made and limited ileocolectomy was done following which ileum was brought outside as temporary ileostomy

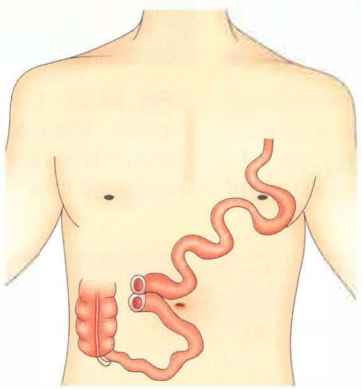


Fig. 28.47: Loop ileostomy is done following resection of perforated enteric ulcer. Distal ileum was friable and oedematous. Hence it is brought out. This is a safer option

CROHN'S DISEASE

It was called regional ileitis because the disease was first reported in the terminal ileum. However, today it is called **regional enteritis** because the disease can occur in jejunum, ileum, colon, oesophagus, etc. Involvement of ileum is more common, followed by colon.

Definition

- Crohn's disease is a chronic transmural inflammatory disease of the gastrointestinal tract of unknown aetiology.

Aetiology

1. **Infectious agents:** *Mycobacterium paratuberculosis* and measles virus have been proposed as potential causes of Crohn's disease. However, it should be noted that antimicrobial therapy has not been effective in eradicating Crohn's (unlike ATT in TB). Also, no immunological reaction has been found.
2. **Immunologic factors:** Similar to UC. Focal ischaemia due to autoimmune reaction has also been considered.
3. **Genetic factors:** Single strongest risk factor for development of Crohn's is a relative with Crohn's disease.
   - Relatively high incidence is found in Ashkenazi jaws.
4. **Smoking:** It increases the risk of Crohn's disease threefold unlike its protective effect against ulcerative colitis.

Pathology (Key Box 28.13)

- Disease **starts in terminal ileum** as ulcerations of intestine in about 60% of cases.
- There is extensive inflammatory oedema and mucosal ulcers are present. Fibrotic thickening of the intestine results in **hose pipe rigidity** of the intestine.
- There are **skip areas** which are characteristic of Crohn's disease (segments of intestine are normal in between).
- **Mesenteric nodes are enlarged.** They can be calcified but do not show any caseation.

KEY BOX 28.13

PATHOLOGY OF CROHN'S DISEASE

Transmural inflammation
Reticulation cobblestone
Aphthoid ulcers
Noncaseating cicatrising granuloma
Skip lesions
Mesenteric lymph nodes enlarged
Ulcers are in ileum, jejunum, colon, etc.
Rigidity—Hose pipe
Adhesions occur soon
Linear ulcers
Remember as **TRANSMURAL**

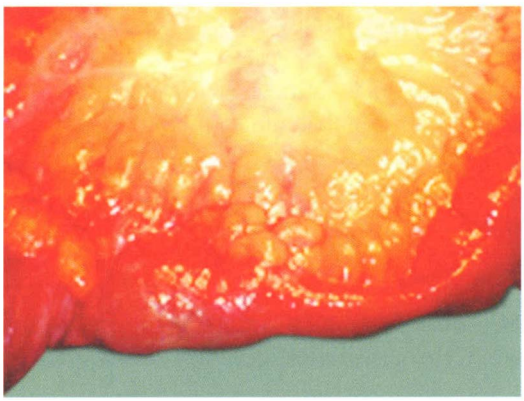


Fig. 28.48: Crohn's disease—creeping fat (*Courtesy:* Dr Ramesh Rajan, Gastrointestinal Surgeon, Trivandrum Medical College, Kerala)

- **Intense infiltration of mononuclear cells** and lymphoid hyperplasia is common.
- As the disease progresses, there is cicatrising granuloma of the bowel wall. This results in narrowing of lumen causing intestinal obstruction. **Caseation is characteristically absent.**
- Once inflammation spreads to the serosa, adhesions develop between bowel loops or other structures. Abscesses occur in the mesentery which rupture resulting in internal fistula (Fig. 28.48).

PEARLS OF WISDOM

Transmural inflammation is characteristic of Crohn's disease.

Clinical features (Key Box 28.14 and Figs 28.49 to 28.51)

The disease is often **insidious**, slowly progressive with a protracted course and commonly affects young adults in the second or third decade of life.

Intermittent colicky lower abdominal pain, diarrhoea and weight loss are common. Depending upon symptoms, it can be classified as follows: (Table 28.4)

1. **Stage of ileocolitis:** Clinically, it presents as pain abdomen and mucus in stools. It is seen in younger patients. It may be associated with fever. Presence of pain and tenderness in the right iliac fossa mimics appendicitis. If there is a mass, it may be confused for an appendicular mass.
2. **Stage of subacute intestinal obstruction** occurs due to stricture of the terminal ileum. Strictures can be multiple. They are not reversible.

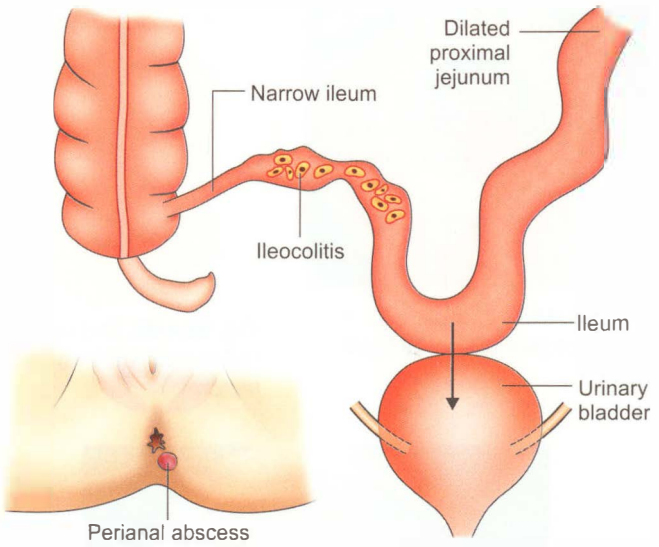


Fig. 28.49: Clinical features of Crohn's disease (*Courtesy:* Ms Vidushi, MBBS student, KMC, Manipal)

KEY BOX 28.14

VARIOUS STAGES/COMPLICATIONS OF CROHN'S DISEASE

Colitis—ileocolitis
Rectal—anorectal disease
Obstruction due to stricture
Hollow viscus fistulae
Nutritional deficiencies
Remember as **CROHN**

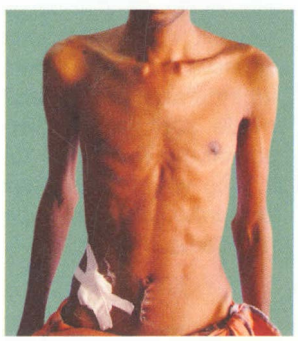


Fig. 28.50: An emaciated patient who underwent appendicectomy, only to present later with features of Crohn's disease

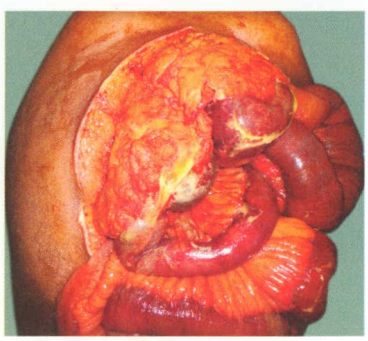


Fig. 28.51: Congested bowel loops, area of perforation, dilated loops of bowel are seen in this picture

Table 28.4 Crohn's disease

Colitis	Obstruction	Mass	Nutrition	Anorectal disease
↓	↓	↓	↓	↓
Diarrhoea	Cramps, distension	Right iliac fossa	Emaciation	Fissure, abscess
↓	↓	↓	↓	↓
Water Weight loss	Vomiting	Gurgling, tender, obstruction	Dehydration	Fistula *in ano*

3. **Stage of fistula formation:** It can be enteroenteric or enterocutaneous. Following are the examples of fistulae encountered in Crohn's disease—ileovesical, ileocolic, ileoileal, ileovaginal, etc.
4. **Perianal disease** in the form of multiple ulcers in the anorectal region, perianal abscesses, multiple fistulae in ano are much more common than in ulcerative colitis. Repeated infection of anal crypt due to diarrhoea is common.

- Extraintestinal complications are similar to ulcerative colitis.

PEARLS OF WISDOM

Anal fissure is the most common anal problem in Crohn's disease. Noncaseating granuloma is most common in anorectal disease.

Differential diagnosis

1. Ulcerative colitis: However, colonic symptoms are more with ulcerative colitis.
2. Tuberculosis: In India, tuberculosis should be considered first and ruled out.
3. Appendicitis: Acute pain in the right iliac fossa can be confused for acute appendicitis. For benefit of the doubt, the patient can undergo laparotomy. Laparoscopy and appendicectomy will be better
4. Intestinal obstruction: Other causes of obstruction have to be kept in mind.

Investigations

1. **Small bowel enema:** Enteroclysis.
   - **Cobblestone** reticulation because of multiple ulcers with islands of normal mucosa in between.
   - Absence of peristalsis in terminal ileum.
   - **String sign of Kantor** is demonstrated in terminal ileum due to narrowing of the lumen.
   - Multiple strictures and dilated segments in between can be demonstrated.
2. **Sigmoidoscopy and colonoscopy** may demonstrate inflamed mucosa, which is granular with **aphthoid ulcers**, which are **discrete** (Fig. 28.52).
3. **Fistulography** to localise the internal fistula.
4. **CT scan:** It is done to detect thickening of bowel and extraintestinal disease.
5. Remember to investigate upper GI tract with gastroduodenoscopy and capsule endoscopy (*see* page 817).

Treatment

I. General principles

- Complete rest, avoid stress and emotions
- A low residue, high caloric diet such as high protein diet
- Some patients may require total parenteral nutrition—some at home called home parenteral nutrition

II. Conservative (medical) treatment is similar to ulcerative colitis.

- Steroids are the mainstay of treatment. They are effective in inducing remission in 70 to 80% of cases.
- Steroids are most effective in treating small intestinal disease. They are anti-inflammatory. They do control diarrhoea, induce remissions. Prednisolone is used for short term treatment. Long-term use can give rise to toxicity such as immunosuppression, bone loss, delayed wound healing, etc.
- Salazopyrines can be used especially in maintenance cases also.
- Even though salazopyrines and corticosteroids have been beneficial in Crohn's disease, salazopyrines do not induce remissions. They are used in acute ileocolitis. Steroids can be used for anorectal disease.
- Immunosuppressive therapy using azathioprine and 6-mercaptopurine are also effective.
- They are effective in treatment of colonic disease. The main concern is bone marrow toxicity. 6-mercaptopurines are known to produce pancreatitis.
- Most recent and promising drug is **infliximab**—a monoclonal antibody to tumour necrosis factor (TNFα). It mainly helps in **closure of fistula**. It is given intravenously and is used for intestinal and perianal disease.
- Metronidazole has shown some benefit.

III. Surgical treatment: Resection is not the aim of surgery but it may have to be done in cases of obstruction, perforation, intra-abdominal abscesses, internal fistulae, bleeding and malignancies. Depending upon the involvement of the bowel, various resections are possible. Examples:

1. Stricture—stricturoplasty or resection
2. Ileocaecal resection (Figs 28.53 and 28.54)
3. Colectomy and ileorectal anastomosis
4. If the fistulae are present, they are disconnected from the bowel and excised.

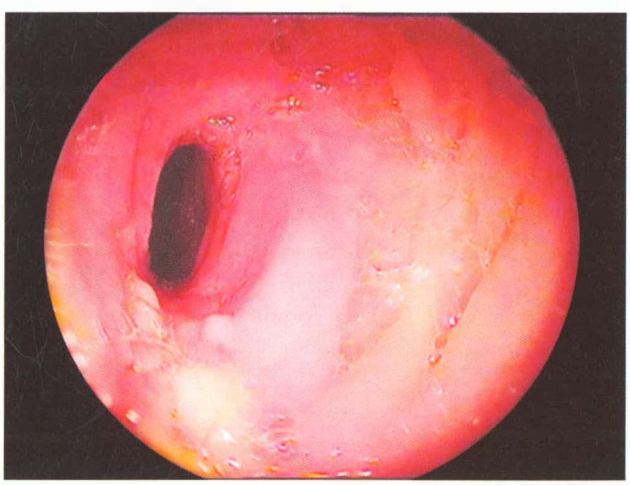


Fig. 28.52: Colonoscopic view of Crohn's disease

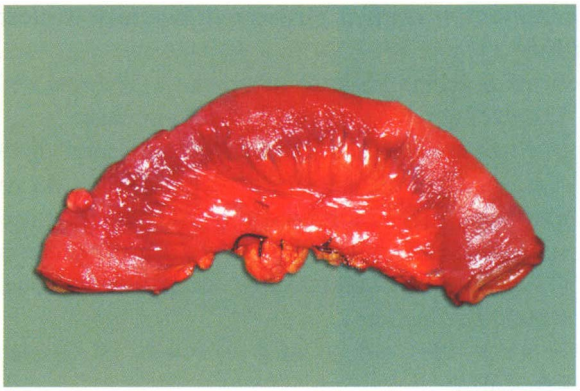


Fig. 28.53: Resected specimen of small intestine. The patient had intestinal obstruction

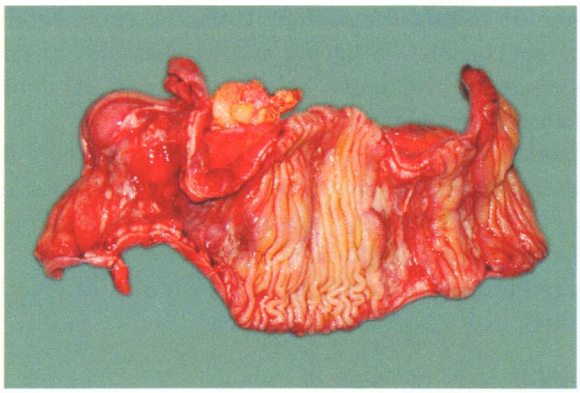


Fig. 28.54: Opened specimen showing stricture. Biopsy proved Crohn's disease

Comparison of intestinal tuberculosis, ulcerative colitis and Crohn's disease is given in Table 28.5.

Prognosis

- In spite of various treatment, there is no cure for the disease. About 10–20% patients come with relapses and recurrent symptoms
- But for repeated treatment including surgical procedures, survival is still good as compared to general population without the disease.

A few observations in crohn's disease

- Ileum is the most common site
- Small bowel alone is affected in 20–30% of the patients
- Both small bowel and large bowel in 50–60% patients
- Duodenum, stomach, oesophagus can also be involved.
- Inflammatory cells and mediators of the inflammation such as cytokines, interleukins, tumour necrosis factors produce the inflammatory changes resulting in granuloma
- Sometimes, very difficult to differentiate clinically and pathologically between tuberculosis and Crohn's disease.
- Anus is involved in Crohn's colitis, not in ulcerative colitis.

SURGICAL COMPLICATIONS OF ENTERIC FEVER

During the third week of enteric fever, *Salmonella typhi*, *paratyphi* (enteric bacilli) multiply in Peyer's patches and can give rise to following problems:

Table 28.5 Summary of tubercular ileocolitis, ulcerative colitis and Crohn's colitis

	Tuberculosis	Crohn's	Ulcerative colitis
1. Bacteria	Mycobacterium	No	No
2. Pathology	Continuous spread. Caseating granuloma, lymph node caseation. Transmural inflammation present	Skip areas. Cobblestone granuloma present. No caseation. Deep longitudinal ulcers. Transmural inflammation	Continuous, pseudopolyposis. Pinpoint ulcers. Crypt abscesses. Superficial ulcers. Mucosa and submucosa involved
3. Site	Ileocaecal	Terminal ileum and colon	Rectum and colon
4. Bowel wall	Thick and fibrotic	Thick fibrotic	Thin
5. **Clinical**			
Bleeding	Uncommon	Uncommon	Very common
Diarrhoea	Uncommon	Very common	Very common
Fever	Common	Common	Rare
Mass	Very common	Common	Rare
Anal disease	Rare	Very common	Rare
Toxic megacolon	Never	Rare	More common
Fistulae	Uncommon	Very common	Uncommon
Stricture	Common	Common	Rare
6. Malignancy	No	Can predispose (low)	High incidence

1. **Haemorrhage** is seen in about 5–10% of cases due to ulceration of Peyer's patches. It can be occult, obvious or rarely massive bleeding. It is managed conservatively in majority of cases.
2. **Perforation of terminal ileum:** An oval, vertical perforation results in peritonitis. Enteric perforation need not give rise to all signs of peritonitis. Guarding and rigidity can be minimal because of poor, immunocompromised nature of the diseases and due to **Zenker's degeneration** of abdominal wall muscles. It is a single perforation in about 85% of the cases. It is situated in the antimesenteric border of the terminal ileum. Typically, it occurs in the third week of enteric fever. Bradycardia, dehydration, toxicity are the other features.
   - **Hyperplasia of reticuloendothelial system** including lymph nodes, liver and spleen is characteristic of typhoid fever (Key Box 28.15).
   - **Diagnosis of perforation** is based clinically on the acute abdominal pain, bleeding per rectum with/without guarding and rigidity. High grade fever, toxicity and bradycardia are other features that help in the diagnosis.
   - **Plain X-ray abdomen** may not reveal gas because of small sealed off perforation.
   - **The most useful investigation is CT scan** which can reveal not only pneumoperitoneum but also pericolic collection, which can be missed by ultrasound (*see* the clinical notes below).

PEARLS OF WISDOM

- ***When you suspect peritonits, no guarding and no rigidity***
- ***When you suspect perforation, no gas under diaphragm***
- ***When you suspect toxicity, no tachycardia but bradycardia, suspect enteric ulcer***

Treatment

- Third generation cephalosporins are used
- **Emergency laparotomy, resection of bowel** and end-to-end anastomosis or closure of the perforation by using non-absorbable sutures. Abdomen is closed with a tube drain kept in the right iliac fossa. Wound infection is common in such cases. Multiple fistulae are also common (Figs 28.55 to 28.58)
- **Small bowel exteriorisation:** This can be considered in cases after resection when both ends of the intestine are friable. This is a very safe option. After 1–4 weeks, relaparotomy and anastomosis of resected ends is done.

3. **Paralytic ileus** due to toxic dilatation of intestine results in distension of abdomen. It is managed by drip and suction.
4. **Typhoid cholecystitis** is not uncommon. Its starts within 2 to 4 days fever. Chances of gallbladder perforation are present.
5. **Typhoid pyelonephritis**, cystitis, epididymo-orchitis.
6. **Typhoid osteomyelitis**
7. **Typhoid conjunctivitis**
8. **Thrombosis** of the common iliac vein occurs probably due to sluggish blood flow.

CLINICAL NOTES

A patient with partially treated enteric fever presented on the 10th day with acute abdominal pain and fever. Lower abdominal guarding and rigidity were present. Plain X-ray abdomen did not reveal gas under the diaphragm. Ultrasonography did not reveal pericolic collection. A CT scan of the abdomen was done. It revealed pneumoperitoneum and pericolic collection. Immediate laparotomy and closure of the perforation was done and peritoneal cavity was drained. The patient recovered dramatically. ***When in doubt, request a CT scan.***

Enteric Ulcer Perforation

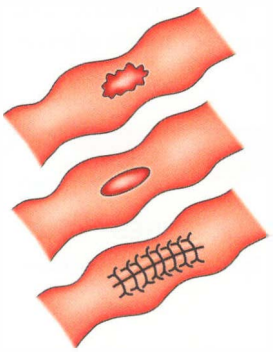


Fig. 28.55: Single ulcer, simple suturing

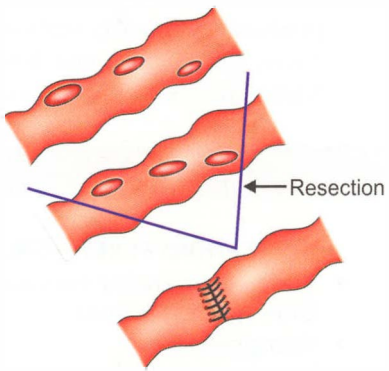


Fig. 28.56: Multiple ulcer—resection and anastomosis

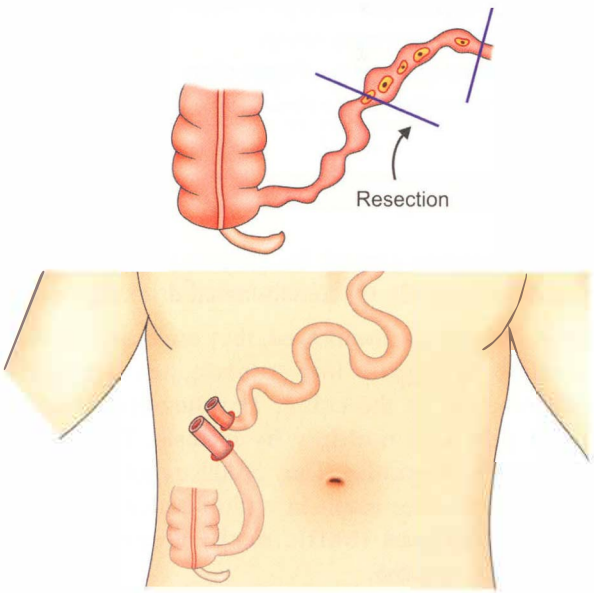


Fig. 28.57: Both ends brought outside – small bowel exteriorisation (*Courtesy:* Dr Vidushi, MBBS student, KMC, Manipal)

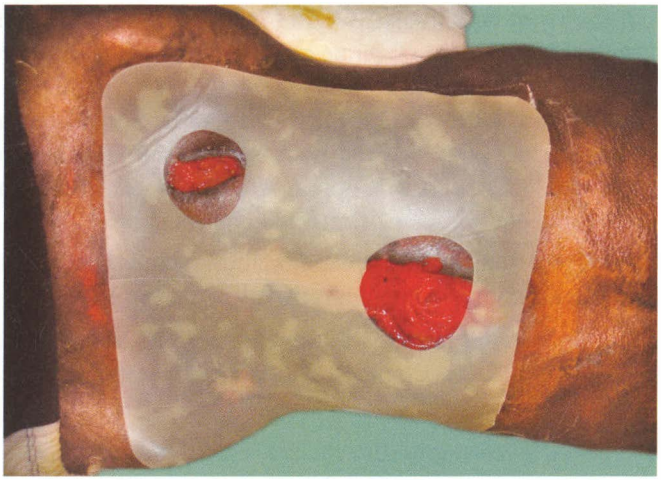


Fig. 28.58: Multiple bud fistulae developed following resection and anastomosis of typhoid ulcers treated with ostomy bags

9. **Perforation** of large intestine can occur in paratyphoid 'B' infections.
   - Most of these complications occur due to bacteraemia produced in the early septicaemic phase of enteric fever. Liver, spleen, bone and bowel are commonly affected. Metastatic abscesses are common.

KEY BOX 28.15

TYPHOID FEVER—SALIENT FEATURES

- **T**oxic—Dehydrated delirious patient with diarrhoea—Green pea soup stools
- **Y**oung patient
- **P**oor tone of the abdominal muscles due to Zenker's degeneration
- **H**igh grade fever
- **O**ther—hepatosplenomegaly, Rose spots
- **I**mmunocompromised status
- **D**ecreased pulse rate—**Faget sign**

You can remember as **TYPHOID**

INTESTINAL AMOEBIASIS

- This disease is caused by *Entamoeba histolytica* and transmitted mainly by contaminated drinking water.
- After the cysts are swallowed, they are broken down in the intestine by trypsin into trophozoites which produce inflammation of the colon. **Trophozoites swallow red blood cells and multiply by mitosis. They enter into crypts of Lieberkuhn.** They produce multiple submucous loculi which later result in multiple ulcers. These ulcers are **flask-shaped (bottle neck) ulcers** with healthy intervening mucosa.
- Some trophozoites are transformed into cysts and excreted outside (Fig. 28.59).

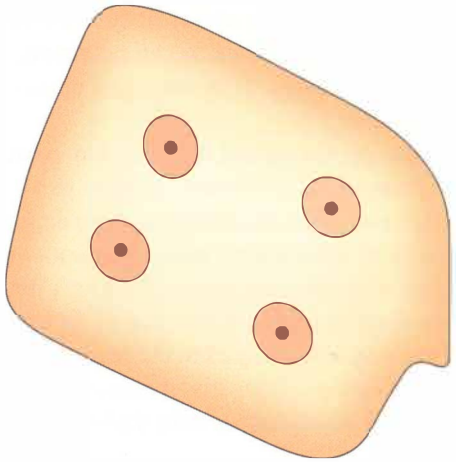


Fig. 28.59: Quadrinucleate cyst of entamoeba

- Lower sigmoid and upper rectum are involved in 75% of cases.

Clinical features

1. **Amoebic typhlitis:** Inflammation of the caecum by amoeba is described as amoebic typhlitis. It produces pain in the right iliac fossa and it can be confused for appendicitis. However, in this condition there is also tenderness in the left iliac fossa. A point on the left side corresponding to McBurney point on the right side is called **Sir Philip Manson-Bahr's amoebic point of tenderness** (Fig. 28.60) and is suggestive of involvement of rectosigmoid.
2. **Amoebic dysentery:** It can be acute or chronic. An acute attack is associated with gripping pain abdomen with blood and mucus in stool, an urgency to pass stools. High grade fever and tenesmus are the other features. Chronic dysentery is more common with 2–4 foul-smelling stools per day and mild to moderate colicky abdominal pain.
3. **Amoeboma:** Chronic, low grade, persistent inflammation of the caecum produces granulomatous hyperplasia of the caecum, with thickening of the pericaecal tissue (producing mass in the right iliac fossa) (Fig. 28.61). Amoeboma can

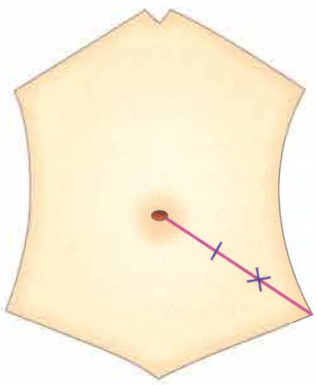


Fig. 28.60: Sir Philip Manson-Bahr's amoebic point

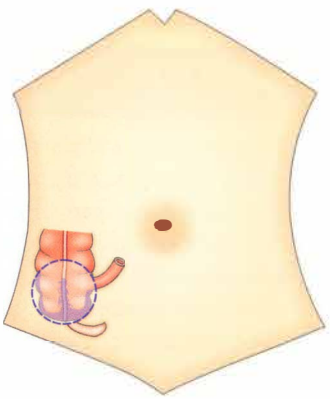


Fig. 28.61: Amoeboma is a tender thickened gurgling mass

also occur in rectosigmoid junction. This is, however, uncommon nowadays because of effective treatment with metronidazole.

- Clinically, this manifests as mass in the right iliac fossa causing dull aching pain, vague ill health, tender palpable caecum with guarding. It can be confused for ileocaecal tuberculosis or carcinoma caecum.
- It responds very well to metronidazole.

4. **Amoebic perforation of caecum** or sigmoid can occur resulting in pericolic abscess. Peritonitis needs emergency surgery.
5. **Massive bleeding per rectum** is rare. It occurs due to separation of the slough.

Treatment

1. **Metronidazole** 400–600 mg, 3 times a day for 10 days. It acts on amoebae present in the lumen and tissue.
2. Diiodohydroxyquin 650 mg, 3 times a day for 20 days is another alternative.
3. Diloxanide furoate is ideal for chronic cases in the dose of 500 mg, 3 times a day for 10 days. It acts on luminal amoebae. It is the drug of choice in chronic cyst passers.
   - Supportive treatment in the form of hospitalisation, correction of dehydration, antispasmodics and bedrest is also advised. Stool culture must be done before and after treatment with antiamoebic drugs.

RADIATION ENTEROPATHY

- These are the changes which occur in the intestines after they are exposed to radiation.
- Typically happen when radiation is given to the pelvis after rectal resections or radiation given to treat carcinoma cervix
- Incidence is 5% after 4500 cGy units to 30% after 6000 cGy units.
- Chief factor is radiation induced damage to the blood vessel supplying the vessel wall.
- **Chronic intestinal ischaemia** occurs resulting in **stricture** and intestinal obstruction. Some cases are due to fibrosis, dense pelvic adhesions, bowel getting entrapped into the fibrosis, fistula formation and pelvic abscess.
- When there is no obstruction, conservative line of treatment to be followed.
- If there is obstruction, laparotomy with resection or bypass or any other treatment depending upon the findings.
- Surgery can be difficult because of dense adhesions.
- Difficult to prevent these changes. However, an **absorbable mesh can be placed** over the small intestines after pelvic surgery when radiation is contemplated.

SMALL BOWEL TUMOURS

- Benign tumours such as lipoma, hamartoma, polyps can occur.
- Malignant tumours such as adenocarcinoma, gastrointestinal stromal tumours **(GIST)**, carcinoid and lymphoma can occur in the small intestine.

PEUTZ-JEGHER SYNDROME (FAMILIAL HAMARTOMATOUS POLYPOSIS)

This syndrome is characterised by:

1. **Familial** tendency

Fig. 28.62: This patient presented to hospital with abdominal pain. The Registrar who examined the case reported that there was a mass abdomen. However, when he presented the case next day, there was no mass. The patient had undergone laparotomy 10 years back for similar complaints. A segment of bowel had been resected earlier. This photograph gives the clue to the diagnosis—pigmentation of lips and oral mucosa. The mass (appearing and disappearing) was due to intussusception (*Courtesy:* Prof Sreevatsa, HOD, Prof Bharathi, Prof Bagli, Dr Srikar Pai, Department of Surgery, MS Ramaiah Medical College, Bengaluru, **CME 2008)**

Table 28.6 Comparison of two hamartomatous polyposis syndromes

Peutz-Jegher syndrome	Cronkhite Canada syndrome
• Hamartomatous polyps	• Hamartomatous polyps
• Polyposis of small intestine (mainly)	• Gastrointestinal polyposis
• Melanin spots on the buccal mucosa and lips	• Cutaneous pigmentation, finger nails atrophy, alopecia
• Bleeding and intussusception are common complaints	• Intestinal obstruction can occur
• Rarely malignancy can occur	• Severe malabsorption—protein-losing enteropathy

CLINICAL NOTES

We had 3 interesting cases of Peutz-Jegher syndrome. ***The first case*** *was of a boy of 14 years, who presented with acute intestinal obstruction. At laparotomy, there were 3 intussusceptions in the jejunum due to polyps. About 15 large polyps were removed after doing an enterotomy.*

The second case *was a 50-year-old lady, who presented with duodenal carcinoma. Endoscopy revealed polyps in the stomach and duodenum. Specimen of pancreatico-duodenectomy revealed it as a case of Peutz-Jegher syndrome. This lady did not have pigmentation of the oral mucosa.*

The third case *was of a 35-year-old male who has been coming to our hospital with intermittent bleeding and anaemia. Endoscopy revealed multiple polyps in the stomach and duodenum. Small bowel enema demonstrated multiple polyps in the small bowel. Even proctoscopy showed multiple polyps in the rectum. He is being managed conservatively.*

2. **Melanosis** of mucosa of lip, cheek, interdigital space and even perianal skin (Fig. 28.62 and Table 28.6).
3. **Multiple polyps** in the small bowel and large bowel mainly in the jejunum. They are hamartomatous polyps.
   - It is an autosomal dominant disease.

Clinical presentation

- Runs in families.
- As a cause of bleeding per rectum, results in chronic anaemia.
- Can cause adult intussusception.
- Rarely, it can turn into malignancy.
- Female patients have increased chances of breast and cervical cancer (*see* clinical notes)

Treatment

- Blood transfusion to correct anaemia
- Resection of that portion of bowel containing polyp, in cases of bleeding or intussusception.

ADENOCARCINOMA

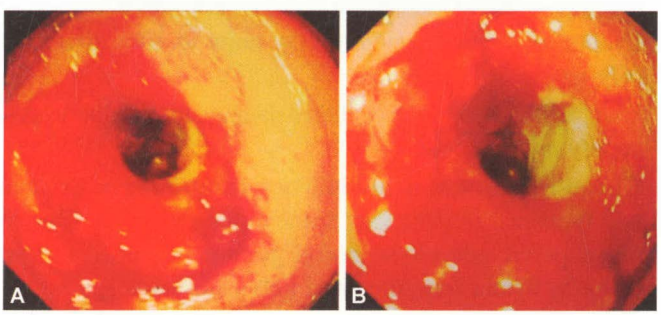
- Incidence is 40% of small bowel tumours. Overall, small bowel tumours are rare. Reasons have been given in Key Box 28.16
- Duodenum is the commonest site of adenocarcinoma. If it arises from first part and second part, it may require Whipple's pancreaticoduodenectomy. From the third part, early cases of adenocarcinoma can be resected without removal of pancreas.
- Some familial diseases predispose to adenocarcinoma. They are familial polyposis coli, Adenomas, Crohn's disease, etc.

KEY BOX 28.16

MALIGNANCY IN SMALL INTESTINE IS RARE—WHY?

- No stasis, rapid transit of food
- Secretion of immunoglobulins—IgA
- Various carcinogens are diluted by secretions
- Comparatively low and inactive bacteria in the small bowel

Fig. 28.63: CECT scan showing jejunal growth with narrowing of the lumen of the jejunum



Figs 28.64A and B: Push enteroscopy showing ulcerative lesion in the proximal jejunum

- Clinical features include vague features like nausea, poor appetite, crampy abdominal pain, bleeding and intestinal obstruction.
- Diagnosis is by CT scan (Fig. 28.63).
- **Push enteroscopy** has the advantage of visualisation of the growth and to take biopsy (Figs 28.64A and B).
- Other option is capsule endoscopy. It is time consuming and biopsy is not possible

Fig. 28.65: Ulcerative lesion at surgery

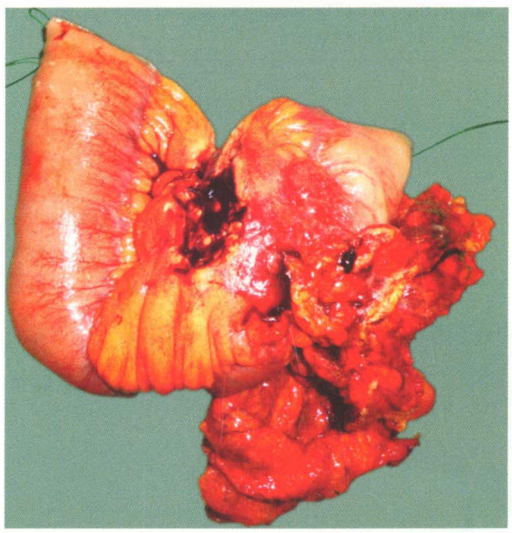


Fig. 28.66: Resection of the growth done with 6 cm margin with lymph nodes

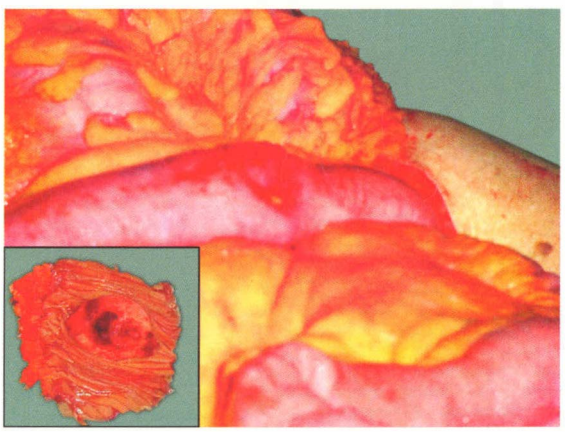


Fig. 28.67: This patient presented with abdominal pain, melaena and loss of weight. CT scan of the abdomen revealed mass in the jejunum. It was resected (inset). Mucosal surface showed ulceration. Histopathology report was adenocarcinoma

- Resection with at least 7 cm margin with lymphadenectomy is the treatment of choice (Figs 28.65 to 28.67).
- Hardly any role for radiotherapy and chemotherapy

GIST: GASTROINTESTINAL STROMAL TUMOUR

- Earlier called as leiomyoma and leiomyosarcoma, they occur in jejunum or ileum. These are uncommon mesenchymal tumours (Figs 28.68 to 28.70).
- **Stomach is the commonest site.** GISTs are rare in the oesophagus whereas leiomyoma is more common.
- They arise from **interstitial cells of Cajal** which are pacemaker cells that regulate motility and peristalsis.
- They present as bleeding/mass/perforation.
- Very often massive bleeding may be the only presentation
- **Carney's triad**
  1. GIST
  2. Pulmonary chondromas
  3. Extra-adrenal paragangliomas.

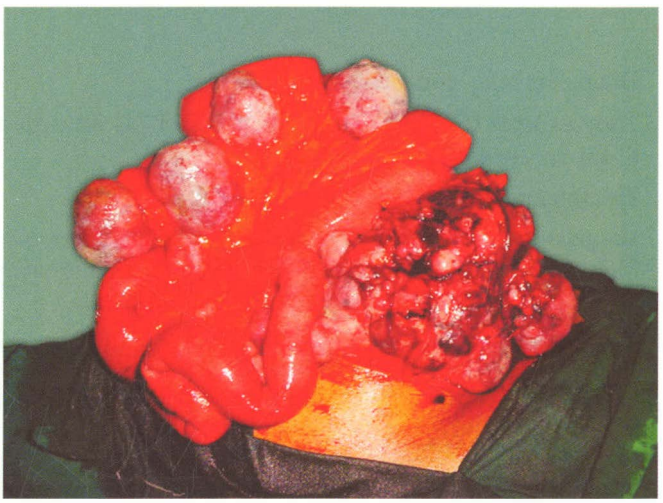


Fig. 28.68: GIST involving terminal ileum, caecum and ascending colon (*Courtesy:* Dr Stanley Mathew, FRCS, Professor, KMC, Manipal)

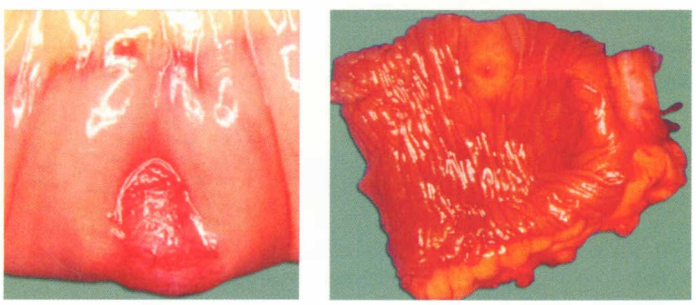


Fig. 28.69: GIST involving jejunum—resected specimen showing mucosal ulceration (*Courtesy:* Dr Padmanabha Bhat, Dept. of Surgery, KMC, Manipal and Prof HD Shenoy, Father Muller's Hospital, Mangalore)

Fig. 28.70: GIST from small intestine (*Courtesy:* Dr Ramesh Rajan, MS, MCh, Surgical Gastroenterologist, Trivandrum, Kerala)

- Diagnosis is by endoscopy and CT scan
- They express CD117/CD34 antigen. **CD117 is known as C-kit receptor.**
- Treatment is in the form of resection.
- Diagnosis of malignancy is by mitotic figures: < 10 mitoses/high power field (HPF) suggests a low grade malignancy.
- > 10 mitoses/HPF suggests high grade malignancy.
- Increased incidence of C-kit has also been found in patients with neurofibroma.

CARCINOID TUMOUR (ARGENTAFFINOMA) AND CARCINOID SYNDROME

It arises from argentaffin/chromaffin cells which are present in the crypts of the villi of intestine. These cells are called as **Kulchitsky cells**. These cells stain with ammonical silver salt solution to black colour. Hence, they are called as argentaffinoma or chromaffinoma. They secrete 5-HT (serotonin or 5-hydroxytryptamine). They can be single or multiple and can be associated with adenocarcinoma.

Sites (Table 28.7)

- *Appendix:* 65%. **It is the most common site.** It occur more commonly in females. When the tumour occurs i the **appendix, it is usually benign,** hard and occurs in dista one-third of appendix (Key Box 28.17).
- **Terminal ileum:** 30%. Most of them are **malignant**. When the tumour occurs in the **ileum it is usually malig nant** and produces multiple bulky secondaries in the liver even when primary is very small. Fibrosis of the mesenter results in kinking of bowel causing periodic abdominal pain

KEY BOX 28.17

'MOST COMMON' FOR CARCINOID APPENDIX

- Most common neoplasm of the appendix
- Mostly found in distal one-third of the appendix
- Mostly tumour is yellow staining for immunohistochemistry
- Mostly benign—very rarely metastasise
- Mostly appendicectomy is sufficient. Rarely, right hemicolectomy may be required.

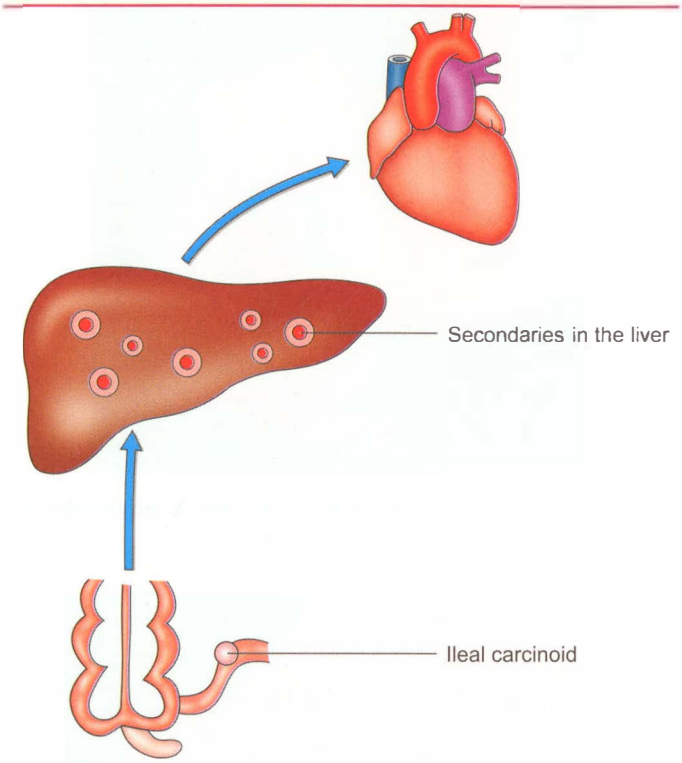


Fig. 28.71: Carcinoid syndrome

Table 28.7 Comparison of carcinoid tumours at various sites

	Foregut	Midgut	Hindgut
Site	Duodenum, stomach, bronchus, pancreas, etc.	Jejunum, ileum, right colon	Rectum
Incidence	Rare	Common	Uncommon
Hormones	No serotonin	5-HT, prostaglandins, insulin, ACTH	Do not secrete

Bronchial carcinoid produces bronchial obstruction and produces carcinoid syndrome without secondaries in liver.

- The hormones produced by the tumour **5-HT** (serotonin) are not metabolised because of the secondaries. So, they are absorbed into the circulation and produce various symptoms. This is called as carcinoid syndrome (Fig. 28.71).
- **Carcinoid syndrome** consists of following features:
  1. Carcinoid tumour which is malignant
  2. Secondaries in the liver
  3. Symptoms are red-blue cyanosis of skin, flushing attack and asthmatic attacks, intestinal hyperperistalsis causing diarrhoea, pulmonary and tricuspid stenosis with CCF.

Diagnosis

- 24-hour urine for 5-hydroxyindole acetic acid. Normal levels 2–9 mg/24 hours. Very high values are found in this condition.
- Ultrasound and CT scan are other investigations which will help in detecting mass lesion or liver metastasis.

Treatment

1. Resection of the tumour with a wide margin along with lymph nodes. Otherwise debulking should be done.
2. **Bromocriptine** 2.5 mg twice a day can be given to reduce the symptoms. Other agents which can be used are methysergide and diphenoxylate hydrochloride.
3. Secondaries in the liver are treated by intra-arterial (hepatic artery) **streptozocin**. Localised liver metastasis can be treated with resection.
4. **Therapeutic embolisation** of hepatic artery by using gel foam, etc. will reduce the size of the liver thereby decreasing discomfort to the patient.
5. Injection octreotide 100 µg IV is the drug of choice in cases of carcinoid crisis (severe bronchospasm).

Octreotide in carcinoid syndrome

1. It is used to suppress the tumour growth
2. Control symptoms – flushing, wheezing, diarrhoea
3. It controls the release of GI hormones
4. Dose is: 100 micrograms subcutaneously tds in patients with mild/moderate, non-life-threatening carcinoid syndrome (Key Box 28.18).

MALIGNANT LYMPHOMA

Primary: Arising from lymphoid tissue
Secondary: Part of systemic lymphoma. **Ileum is the most common site of lymphoma** (Key Box 28.19).

Precipitating factors

1. Crohn's disease
2. Coeliac disease
3. Immunosuppression—AIDS—Usually it is a non-Hodgkin's lymphoma of 'B' cell origin.

KEY BOX 28.18

CARCINOID TUMOURS

- They are APUDOMAS.
- Arise from enterochromaffin cells.
- They may be associated with multiple endocrine neoplasia (MEN) type 1 and 2
- About 15% of carcinoid patients may have carcinoma elsewhere, example: carcinoma stomach, carcinoma lung or carcinoma colon.
- Common age is 25–45 years of age
- Majority arise from midgut—almost 85% cases. They secrete 5-HT
- Appendix is the most common site. Often appendicectomy is enough for malignant carcinoid tumour.
- Foregut carcinoids do not secrete serotonin
- Flushing and diarrhoea can be controlled by octreotide.
- Secondaries can be multiple and big, but slow growing like primary. Arterial embolisation has been tried in many cases.
- Since they are slow growing and prognosis is good-aggressive treatment in the form of surgery, octreotide, liver resections and rarely liver transplantation has been done

KEY BOX 28.19

MALIGNANT LYMPHOMA

1. Western type

- Non-Hodgkin's, B-cell type
- Annular ulcerating lesion
- Bleeding, obstruction, perforation and weight loss

2. Primary lymphoma with coeliac disease

- Increased incidence of lymphoma
- It is a T-cell lympoma
- Diarrhoea, pyrexia are other features

3. Mediterranean—associated with alpha chain disease.[1]

[1]Alpha chain disease is a disorder characterised by the secretion of a defective α-heavy chain. Patients present with steatorrhoea, often progressive and fatal.

KEY BOX 28.20

SHORT GUT—CAUSES

- Mesenteric infarction
- Midgut volvulus
- Necrotising enteritis
- Crohn's disease[1]
- Radiation enteritis

[1]It is common cause in western countries

SHORT GUT SYNDROME

Causes of short gut syndrome (Key Box 28.20)

- Short gut syndrome occurs due to **massive resection** of the bowel resulting in loss of length of the bowel, loss of absorptive area of the bowel and loss of valves. Superior mesenteric artery being an end artery, thrombosis at its origin is invariably fatal.
- **Midgut volvulus** of neonates is congenital due to arrested rotation resulting in floating caecum and mobile intestine. We had an interesting case of midgut volvulus in an 18-year-old boy consequent to a laparotomy done for perforated duodenal ulcer. While replacing the coils of bowel within the abdomen, the mesentery was probably twisted resulting in massive gangrene. This boy now has about 100 cm of the small bowel and surviving on "baby food" for the last 5 years.
- **Necrotising enteritis** (enteritis necroticans) is a complication of infection of small bowel by *Clostridium perfringens*. It usually occurs after a heavy feast where pork is consumed. There is extensive suppuration of mucosal and submucosal layer of jejunum (also ileum). Serosa may show multiple dark bluish patches. Massive resection is done for a necrotic, perforated, unhealthy bowel which results in short gut syndrome.
- **Radiation enteritis** or radiation enteropathy results in patients who receive radiotherapy to the abdominal and pelvic regions, e.g. carcinoma cervix. Arrest of cell division resulting in mucosal thinning, ulceration followed by oedema and later, fibrosis are characteristics of this condition. Endarteritis and vasculitis also add to these changes resulting in stricture, perforations, abscess, mal absorption and multiple resection, etc.

Pathophysiological effects
(Fig. 28.72 and Key Box 28.21)

It depends upon:

- Extent of resection
- Site of resection
- Presence/absence of ileocaecal valve
- Age of the patients
- Infants tolerate extensive resections better than adults. Patients with less than 100 cm of the small bowel will develop severe nutritional deficiencies and may require parenteral nutrition.

KEY BOX 28.21

EFFECTS OF SHORT GUT

- Severe malabsorption
- Gall stones
- Fatty infiltration of liver
- Urinary stones
- Gastric hyperacidity

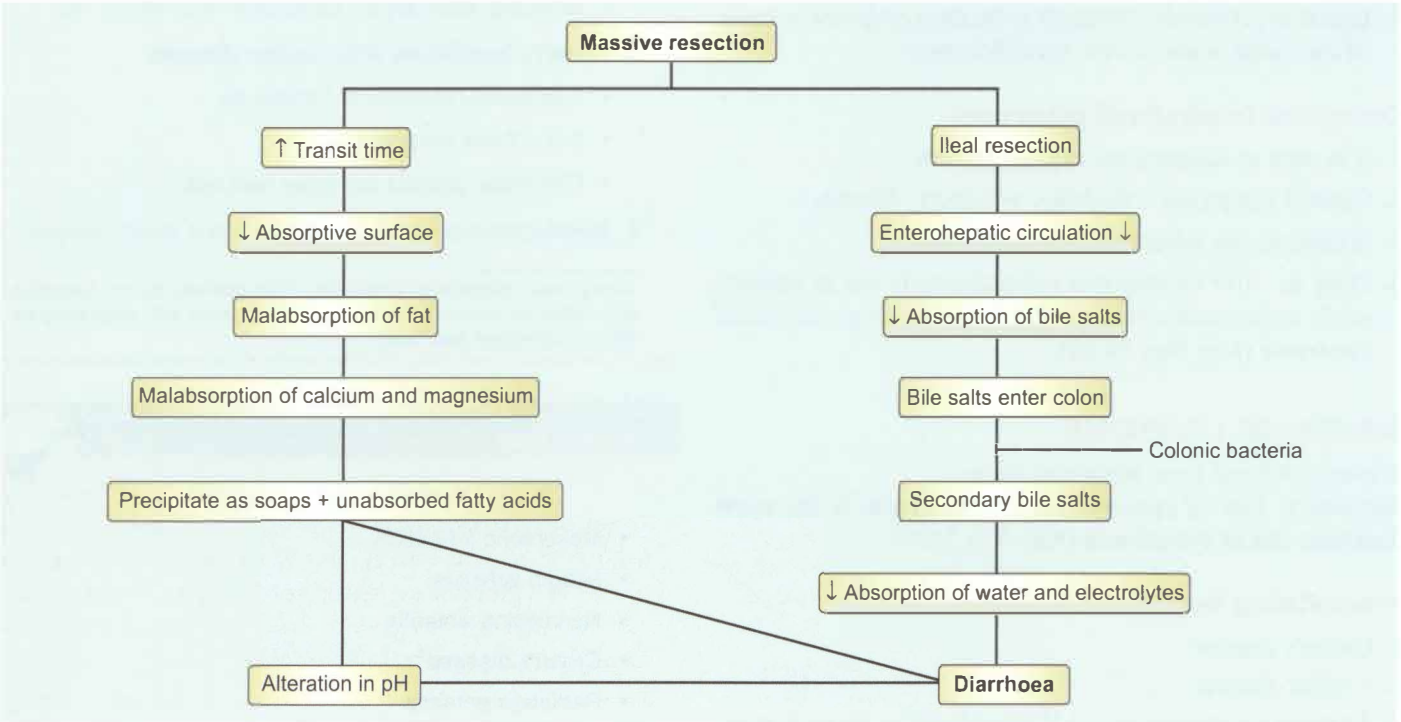


Fig. 28.72: Nutritional deficiency in short gut syndrome

1. **Malabsorption of fat and fat soluble vitamins:** Can occur after ileal resections due to interruption of enterohepatic circulation of bile salts. These bile salts enter the colon and are converted into secondary bile salts. These bile salts block absorption of water and electrolytes.
2. **Gastric hypersecretion:** Due to delayed clearance of gastrin, as in proximal jejunal resections, there is increased gastric secretion of acid resulting in hyperacidity.
3. **Liver disease:** Fatty infiltration of the liver and mild hyperbilirubinaemia is a feature in massive resections and jejunoileal bypass. Acute fulminant hepatic failure can also occur.
4. **Gall stone formation:** There is increased incidence of cholesterol stones as a result of reduced bile salt pool, after ileal resection and jejunoileal bypass.
5. **Urinary stones**
   - All types of urinary stones are common due to low levels of calcium excretion in the urine and high levels of oxalate.
   - Water and salt depletion, and loss of K^+ causes hyponatraemia and hypokalaemia.

Adaptation (Key Box 28.22)

- As a result of loss of significant bowel, dilatation of the remaining intestine and villous enlargement takes place. This is brought about by a humoral agent, enteroglucagon. In children, length of the bowel is increased. Also, the number of cells in the villi is increased (work hypertrophy). There is also evidence to suggest gradual slowing of the transit time.

KEY BOX 28.22

ADAPTATION TO SHORT GUT SYNDROME

- Villus—size
- Length of bowel
- Transit time
- Absorption from colon

Treatment

- Treatment of short gut patients is difficult. It needs a special set up of dieticians who plan "proper food" for these patients in consultation with treating surgeons. It is a gradual process of feeding the patient beginning with parenteral nutrition and progressing to normal, low fat diet after a few months.
- In the initial 2–3 months following massive resection, total parenteral nutrition including supplementation of fluid and electrolytes is the ideal treatment. Sips of plain water or oral hypotonic solutions can be allowed.
- After 2–3 months, when adaptation of bowel takes place, enteral feeding is started gradually with baby food, fat-free, fibre-free, protein rich, liquid diet. Essential fatty acids should be supplied. Diarrhoea is a common problem and is treated with loperamide tablets.
- Enteral feeding can contain low fatty diet in addition to the other nutrients mentioned above.
- **They occur in the mesenteric border unlike Meckel's which occur in the antemesenteric border.**

INTESTINAL FISTULAE

Introduction

- Intestinal fistulae are **abnormal communications** between two portions of the intestine, between the intestine and some other hollow viscus, or between the intestine and the skin of the abdominal wall.
- When it involves **skin and intestines, it is called enterocutaneous fistula.**
- Despite significant advances in their management, intestinal fistulae remain a major clinical problem, with an overall mortality rate of 15 to 25%.

Classification

Anatomical

- Internal: Colovesical fistula
- External: Duodenal, jejunal fistula
- Mixed: Crohn's disease

Depending on the contents

• Low output	< 200 ml
• Moderate output	200–500 ml
• High output	> 500 ml

Aetiology

- **Iatrogenic (70%)—postoperative:** Injury to intestines unnoticed at the time of surgery or injury recognised at the time of surgery, sutured but has given way are prime causes of enterocutaneous fistula. **Anastomotic leak, partial or complete, is another important cause of enterocutaneous fistula. Meticulous surgical techniques, gentle handling of the bowel, usage of the proper suture material (Vicryl and silk), ensuring adequate blood** supply to the intestines which have to be anastomosed will largely prevent anastomotic leak (Fig. 28.73).
- **Stump blow out: Duodenal blow out: Typically happens 4–5 days following surgery** (*see* page 506).

Fig. 28.73: Small intestinal fistulae due to anastomotic disruption and wound dehiscence

- Inadequate resection of the **diseased segment:** In such situations, anastomosis may be in unhealthy bowel.
- **Instrumentation:** Oesophageal perforations are dangerous ones and can occur even with flexible scopies.
- **Spontaneous (30%):** Crohn's disease, diverticular disease of the colon, appendicitis are also known to give rise to fistulae.

Local factors precluding spontaneous closure

- **B**leeding
- **E**nd fistula: Bowel discontinuity
- **F**oreign body: Swabs, tips of the suction tubes (detached), retained sponges or instruments.
- **R**adiation
- **I**nflammation/infection/inflammatory bowel disease
- **E**pithelialisation of the tract is another important cause of the persistent fistula.
- **N**eoplasms

CLINICAL NOTES

A 60-year-old man was operated for appendicitis. On the fifth postoperative day, he developed faecal discharge from the wound. Conservative management was done for 7 days with total parenteral nutrition and other measures. Fistula persisted. CT scan was done on the 14th day. It revealed a ***growth in the hepatic flexure almost encircling the lumen.*** *Colonoscopy and biopsy proved adenocarcinoma colon. He underwent re-exploration and right hemicolectomy was done. The fistula stopped and he was discharged from the hospital after 10 days. This case highlights the fact that in the presence of a distal obstruction, fistula will not close. More importantly, what the patient had was not appendicitis but subacute intestinal obstruction ! The pain was due to colic.*

- **D**istal obstruction
- **Multiple fistulae** that do not heal as in Crohn's or typhoic perforations.
- **Lateral duodenal fistula** does not heal because large volume of contents are poured into the second part of duodenum and there is no rest to the part.
- When the **defect is more than 1 cm**, and the **track is more than 2 cm**, fistula does not close (*see* clinical notes).

Management: Principles

It can be discussed under the following headings.

1. Recognition and aetiology
2. Phase of stabilisation
3. Nutrition–more details on page 207
4. Investigative phase
5. Phase of definitive management
   a. Surgery
   b. Skin care
   c. Abdominal wall defect

1. Recognition and aetiology

- Delay in recovery from paralytic ileus or a common phrase used—when the ***patient is not doing well*** are early indications of a leak or a breakdown of anastomosis. Once the fistula establishes, diagnosis is easy because the drain will start draining intestinal contents including food particles (if oral intake is started). Intra-abdominal collections, abscess, fever, sepsis are other features of leak.
- Recognising the cause is important because it can dictate treatment. To give an example: **If any foreign body is left in the abdomen, exploration and removal may be urgently required.**
- If a specific disease is suspected such as tuberculosis, anti-tuberculous treatment may cure a fistula.

2. Phase of stabilisation

- Nil by mouth, total bowel rest, introduction of a nasogastric tube (Ryle's tube) with continuous drainage decreases fistula output.
- Proton pump inhibitors decrease gastric secretions.
- Protection of skin is by liberal zinc oxide application.
- Common fluid and electrolyte problems seen in patients with GI fistula include dehydration, hyponatraemia, hypokalaemia and metabolic acidosis. They have to be corrected.
- Drainage of collections by CT or ultrasound-guided aspiration or pig tail catheter insertions.
- Broad-spectrum antibiotics, with anaerobic coverage.

3. Nutrition (more details on page 207)

- Nutrition has been discussed in detail in the chapter on fluid-electrolyte balance and nutrition. A few points worth

mentioning are that more distal the fistula, less is the requirement of **total parenteral nutrition** (TPN). TPN is safe today. One may have to wait for 4–6 weeks with TPN for the final repair of complicated fistula such as lateral duodenal fistula or ileal fistula.

- Placement of **nasogastric, nasojejunal tubes, percutaneous gastrostomy or jejunostomy** for nutrition is an important step which can be done with the help of radiology and imaging department.

4. Investigative phase

a. Routine biochemical and haematological investigations

- Blood urea, serum creatinine, blood sugar
- Serum electrolytes
- Serum albumin, transferrin
- Blood culture helps in giving appropriate antibiotics
- Chest X-ray to rule out static pneumonia or ARDS, pleural effusions (source of sepsis also).

b. Imaging studies

- **USG:** Drainage of collections by catheters. If drainage persists, suspect fistulous communication.
- **Fistulograms** to define the exact site of fistula—proximal or distal—gastric or intestinal (Key Box 28.23).

KEY BOX 28.23

FISTULOGRAM/CT/CONTRAST GI SERIES

- What is the cause of the fistula?
- Is the bowel completely disrupted or is it a lateral fistula with the bowel in continuity?
- What is the length of the fistula tract?
- Is there an abscess cavity?
- What is the size of the bowel wall defect?
- Is there a distal obstruction?

- **Contrast CT abdomen:** To detect collections, discontinuity, diseased segments, distal obstruction foreign body. Extremely useful investigation in intestinal fistula.
- **Colonoscopy:** Barium enema are other investigations used in specific situations such as colonic fistulae.

5. Phase of definitive management

a. Surgical intervention

- **Drainage or aspiration of pus** urgently when there is sepsis before doing an elective repair.
- **Feeding jejunostomy** as in high fistulae, e.g. duodenal or oesophageal fistula.
- **Diversion/exclusion/colostomy/bypass** are various other treatments depending upon the location of fistula (Fig. 28.74).
- **Definitive treatment**: It includes restoration of continuity-resection and anastomosis, internal diversion or exteriorisation of bowel as in suspicious viability (mesenteric ischaemia or enteric perforations—friable bowel, known for leak and releak).

Fig. 28.74: Small bowel exteriorisation was done for enteric perforation after resection—safe option when there is gross contamination

Fig. 28.75: Severe nutritional deficiency following duodenal fistula—autocannibalism

Fig. 28.76: Extensive excoriation of the skin in a faecal fistula. Zinc oxide has been applied to the abdominal wall

b. Methods of skin care (Figs 28.75 and 28.76)

- Pouches, stoma bags of different sizes are available.
- Creams and ointments—zinc or petroleum-based
- Suction catheter placed *in situ*: Low pressure suction 60–80 mmHg.

Generally, dressings need to be changed every 4th hourly. Pouch/reservoir system should be employed.

c. Abdominal wall defects

- Depending upon the size of the defect, immediate or delayed closure can be achieved with tension-releasing incisions on the external oblique or laparostomy and secondary suturing, skin grafting or by using prosthetic mesh such as Marlex or Prolene.

SMALL INTESTINAL DIVERTICULA

Introduction

Small intestinal diverticula are far less common than colonic diverticula. Multiple sac-like mucosal herniations occur

through weak points in the intestinal wall where blood vessels penetrate.

Incidence

Diverticula are more common in duodenum than jejunum or ileum (Fig. 28.77).

Pathophysiology

- It is believed to develop as a result of abnormalities in peristalsis, intestinal dyskinesis and high segmental intraluminal pressures.
- **They occur in the mesenteric border unlike Meckel's which occurs in the antimesenteric border.**
- Diet low in fibre and rich in fat and some visceral myopathy have been blamed for development of diverticula.

Clinical

- Asymptomatic
- Occult blood in the stools—positive
- Anaemia and symptoms such as fatigue, weakness, peda oedema.
- Malabsorption—diarrhoea, flatulence, weight loss
- Pain abdomen, signs of peritonitis such as tenderness guarding or localised abscess.

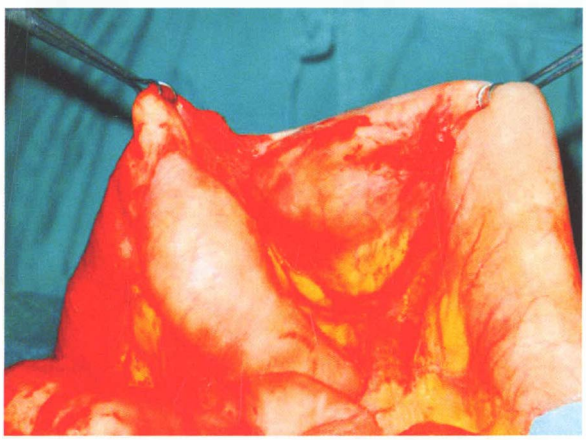
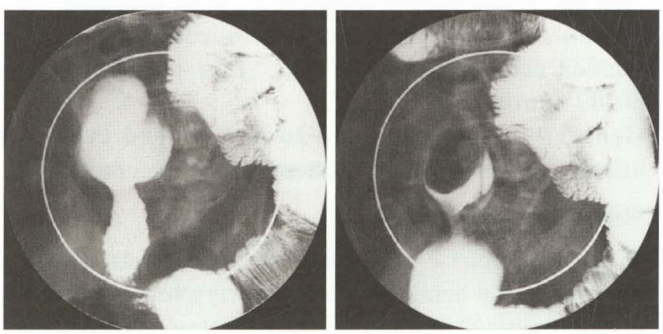


Fig. 28.77: Diverticula form mesenteric border of jejunum. It was the cause of anaemia due to occult blood loss



Figs 28.78 and 28.79: Enteroclysis. First picture showing filling defect and a clear view of diverticula in the second picture

Investigations

- Specific investigations are enteroclysis and enteroscopy.
- CT with contrast is the ideal investigation.
- Often diverticula are not suspected but on a CT scan obtainec for some pathology in the abdomen may reveal a localisec abscess. Later, at laparotomy it can turn out to be diverticula (Figs 28.78 and 28.79).

Treatment

- Resection and anastomosis of the bowel containing diverticula.

INTERESTING 'MOST COMMON' FOR INTESTINES

- Most common site of gastrointestinal tuberculosis is ileocaecal region.
- Most common site of ulcerative colitis is rectum.
- Most common extraintestinal manifestations of ulcerative colitis are peripheral arthritis and ankylosing spondylitis.
- Most common site of gastrointestinal perforation in enteric fever is terminal ileum.
- Most common neoplastic lesion of small intestine is lymphoma.
- Most common anal problem in Crohn's disease is fissure in ano.
- Most common site of adenocarcinoma in intestines is duodenum.
- Most common site of malignant carcinoids is terminal ileum.
- Most common site of carcinoid in intestines is appendix.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated with flowcharts and coloured pictures.
- Small intestinal divericula, intestinal fistulae and tumours have been discussed in more detail.
- Radiation enteropathy has been added.
- Multiple choice questions have been added

MULTIPLE CHOICE QUESTIONS

1. Pathology of ulcerative colitis includes all of the following *except:*
A. Punched out ulcers
B. Pseudopolyposis
C. Pipe stem colon
D. Pus cells

2. Toxic megacolon is seen in the following *except:*
A. Intestinal tuberculosis
B. Ulcerative colitis
C. Amoebic colitis
D. Salmonella colitis

3. String sign of Kantor is seen in:
A. Crohn's disease
B. Tubercular enteritis
C. Typhoid enteritis
D. Amoebic colitis

4. Guarding and rigidity can be minimal in enteric perforation of terminal ileum because:
A. The perforation is usually small
B. It is self-sealing and self-limiting
C. Zenker's degeneration of abdominal muscles
D. Occurs in the antimesenteric border of the terminal ileum

5. Flask-shaped (bottle neck) ulcers are a feature of:
A. Intestinal tuberculosis
B. Crohn's disease
C. Intestinal amoebiasis
D. Typhoid enteritis

6. Malignancy in small intestine is rare because:
A. Bacteria in the intestine are protective
B. Blood supply is very good
C. No stasis, rapid transit of food
D. Low levels of immunoglubulins

7. Most common site of gastrointestinal perforation in enteric fever is:
A. Duodenum
B. Jejunum
C. Colon
D. Terminal ileum

8. Adaptation to short gut syndrome includes all of the following *except*:
A. Decreased transit time
B. Increased villus size
C. Absorption from colon
D. Increased length of bowel

9. The symptoms of carcinoid syndrome is due to:
A. Histamine B. Serotonin
C. Prostaglandins D. Epinephrine

10. Tenderness in the Sir Philip Manson-Bahr's point is a feature of:
A. Rectosigmoid involvement in amaoebic colitis
B. Acute appendicitis
C. Acute cholecystitis
D. Crohn's disease

11. The commonest site for gastrointestinal stromal tumour is:
A. Oesophagus B. Stomach
C. Duodenum D. Jejunum

12. Carney's triad includes all of the following *except:*
A. Gastrointestinal stromal tumours
B. Pulmonary chondromas
C. Extra-adrenal chondromas
D. Familial hamartomatous polyposis

13. One of the drugs used in the treatment of carcinoid syndrome is:
A. Metronidazole B. Bromocriptine
C. Streptomycin D. Serotonin

14. The most common site for a carcinoid tumour is:
A. Stomach B. Duodenum
C. Appendix D. Rectum

15. Causes of short gut syndrome include all of the following *except*:
A. Mesenteric ischaemia
B. Necrotising enterocolitis
C. Crohn's disease
D. Radiation enteritis

16. C-kit receptor is expressed in:
A. Gastrointestinal stromal tumour
B. Peutz-Jegher syndrome
C. Carcinoid syndrome
D. Crohn's syndrome

17. The most recent and promising drug used in the treatment of Crohn's disease is:
A. Trastuzumab B. Infliximab
C. Monteleukast D. Fab antibodies

18. Most common anal problem in Crohn's disease is:
A. Anal fistula B. Haemorrhoids
C. Anal fissure D. Perianal abscess

19. Transmural inflammation is characteristic of:
A. Crohn's disease B. Tuberculous enteritis
C. Typhoid enteritis D. Ulcerative colitis

20. 'Hose pipe rigidity' is a feature of:
A. Crohn's disease B. Tuberculous enteritis
C. Typhoid enteritis D. Ulcerative colitis

ANSWERS

1 A	2 A	3 A	4 C	5 C	6 C	7 D	8 A	9 B	10 A
11 B	12 D	13 B	14 C	15 D	16 A	17 B	18 C	19 A	20 A

29

Large Intestine

- Surgical anatomy
- Function
- Tumours
- Familial polyposis coli
- Hereditary nonpolyposis
- Colorectal cancer
- Carcinoma
- Colon screening
- Diverticular disease
- Faecal fistula
- Colonic stricture
- What is new?/Recent advances

SURGICAL ANATOMY

- Large intestine extends from ileocaecal valve to anus. It has five segments: right colon, left colon, transverse colon, sigmoid colon, rectum and anal canal (Fig. 29.1).
- Average length is about 135–150 cm.
- Interestingly, alternating portions of the colon are mobile and fixed. Ascending colon and descending colon are fixed but caecum, transverse colon and sigmoid colon are mobile. Mobile structures can undergo twisting (volvulus).
- Layers of colon: Mucosa, submucosa, muscularis propria and serosa. Inner circular and outer longitudinal muscle layer constitute muscularis propria. Inspite of 4 layers, wall of the colon is thin. Hence it distends much more in obstruction.

Caecum

- 7.5 cm in both length and breadth
- Blind pouch
- Completely covered by peritoneum
- Mobile

Diseases

- Carcinoma
- Tuberculosis
- Volvulus—rare
- Amoebic typhlitis
- Intussusception

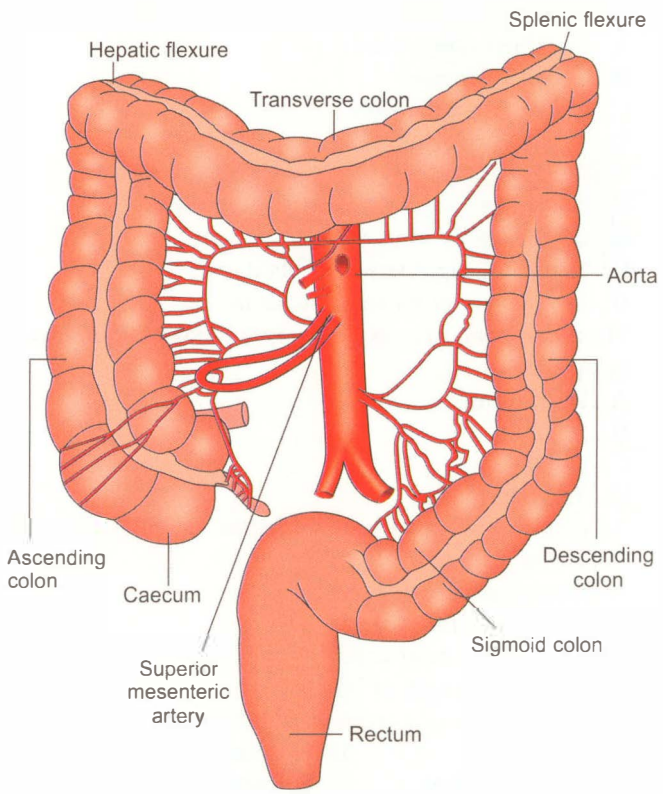


Fig. 29.1: Anatomy of the colon and parts

Ascending colon—15 cm long

- Caecum continues as ascending colon.
- Covered by peritoneum in front and on both sides.
- In 25% of patients, it has a mesentery.

- Right paracolic gutter is deep on the lateral aspect of ascending colon—space for para-colic abscess in cases of perforation peritonitis.

Transverse colon—50 cm long

- Most mobile part of large intestine
- It is suspended by transverse mesocolon, loops down and is adherent to the posterior wall of omental bursa.

Diseases

- Cancer
- Ulcerative colitis

Descending colon—25 cm long

- Continues as sigmoid colon
- Retroperitoneal (like ascending colon)
- Also has paracolic gutter

Sigmoid—40 cm long

- S-shaped
- Ends as rectosigmoid junction where taenia coli ends.

Diseases

- Volvulus
- Diverticulosis
- Cancer

Right colon (Table 29.1)

Big and hepatic flexure is broad.

Left colon

- Small and splenic flexure is acute. Hence, **ischaemic colitis commonly affects splenic flexure** (Table 29.1).
- Splenic flexure is deeply situated. Therefore, malignancy in this area can be easily missed.

Muscle coat

Outer longitudinal muscle is arranged in the form of **three strips** called **taenia coli.** All three join at the rectosigmoid junction and form a complete longitudinal layer of the rectum. **These three taenia coli converge at the base of the appendix.** This is an important method to localise appendix. Inner circular muscle coalesces distally to form internal anal sphincter.

Recognise large intestine by

1. Taenia coli
2. Omental appendices—appendicular epiploicae
3. Haustrations
4. Large diameter (calibre)

Names of three taenia coli

1. Mesocolic: Transverse and sigmoid colon are attached by this.
2. Omental: To which omental appendices attach.
3. Libra (free): Nothing is attached.

PEARLS OF WISDOM

Since taenia are shorter than intestine, the colon becomes sacculated between taenia forming haustra.

Arterial supply (Fig. 29.2)

1. **Superior mesenteric artery**, a branch of abdominal aorta arises at the level of first lumbar vertebra (L1). It supplies the entire small bowel and the right colon up to proximal 2/3rds of transverse colon. **Branches** of superior mesenteric artery (SMA) supplying colon are:

   A. **Middle colic artery** supplies ascending colon, hepatic flexure and the transverse colon mainly. It divides into right and left branches.

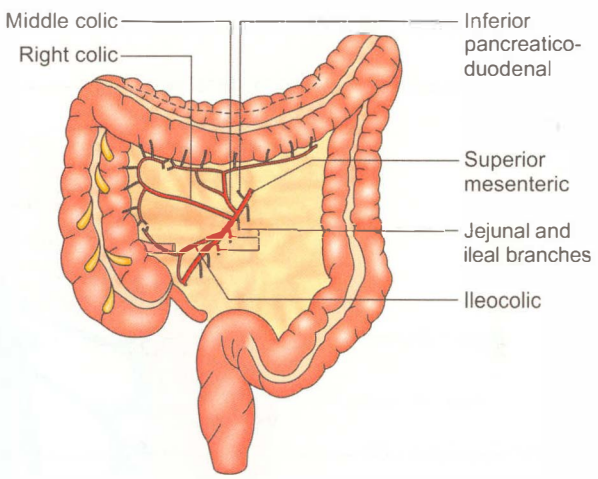


Fig. 29.2: Blood supply of the right colon

Table 29.1 Comparison between right colon and left colon

Right colon	Left colon
1. Big	1. Small
2. Hepatic flexure is broad	2. Splenic flexure is acute and narrow
3. Hepatic flexure is not deep	3. Splenic flexure is deep
4. Paracolic space is broad	4. Left paracolic space is narrow
5. Luminal diameter is big	5. Luminal diameter is narrow
6. Contents are liquid	6. Contents are solid
7. Growth pattern is usually ulceroproliferative	7. Growth is annular stricture
8. Obstruction is not common	8. Hence, obstruction is common
9. More common in women	9. More common in men

B. **Right colic artery** supplies right colon.
C. **Ileocolic artery** supplies the terminal ileum and ascending colon. It divides into anterior and posterior caecal branches and supplies caecum and appendix through appendicular artery.

2. **Inferior mesenteric artery (IMA)**, a branch of abdominal aorta arising at the level of L3 supplies the left colon up to mucocutaneous junction at the lower end of anal canal (Hilton's line). Its branches are (Fig. 29.3):
   A. **Left colic artery** which anastomoses with branches of middle colic artery. It divides into upper and lower branches supplying the descending colon.
   B. **Three sigmoidal branches** supply sigmoid colon
   C. **Superior haemorrhoidal artery** (rectal)

- The anastomotic branches form **marginal artery of Drummond,** which is relatively narrow in the region of splenic flexure (another reason for development of ischaemia).
- **Arc of Riolan** is the anastomotic arcade formed between branches of IMA and SMA.

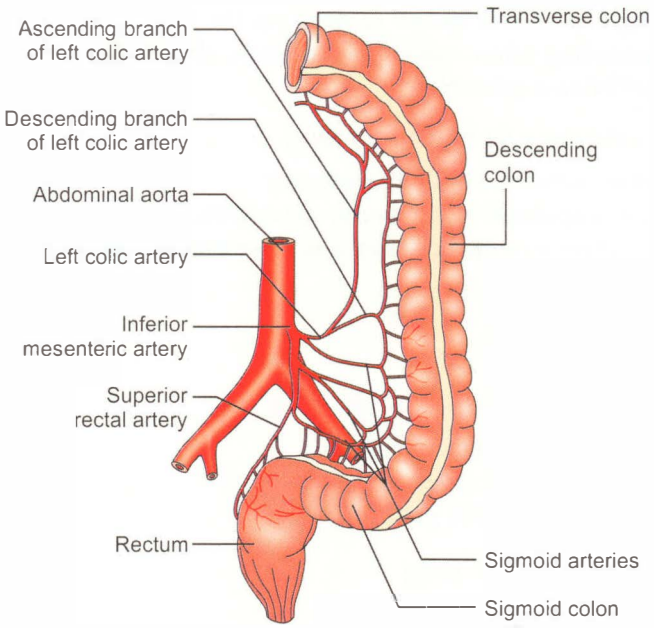


Fig. 29.3: Arterial supply of left colon

Venous supply

- Follows the corresponding artery and empty into superior and inferior mesenteric veins, ultimately draining into the portal vein (Fig. 29.4).
- Thus, any inflammatory conditions of the colorectal/ surgical procedures if they infected, infection can easily spread into portal vein and result in portal pyaemia.

Lymphatic drainage (Fig. 29.5)

- **N1:** Epicolic, paracolic nodes are the first to get involved.
- **N2:** Nodes at the origin of ileocolic and middle colic arteries—intermediate nodes.
- **N3:** Nodes at the origin of superior and inferior mesenteric arteries. They are involved in approximately 50% of the

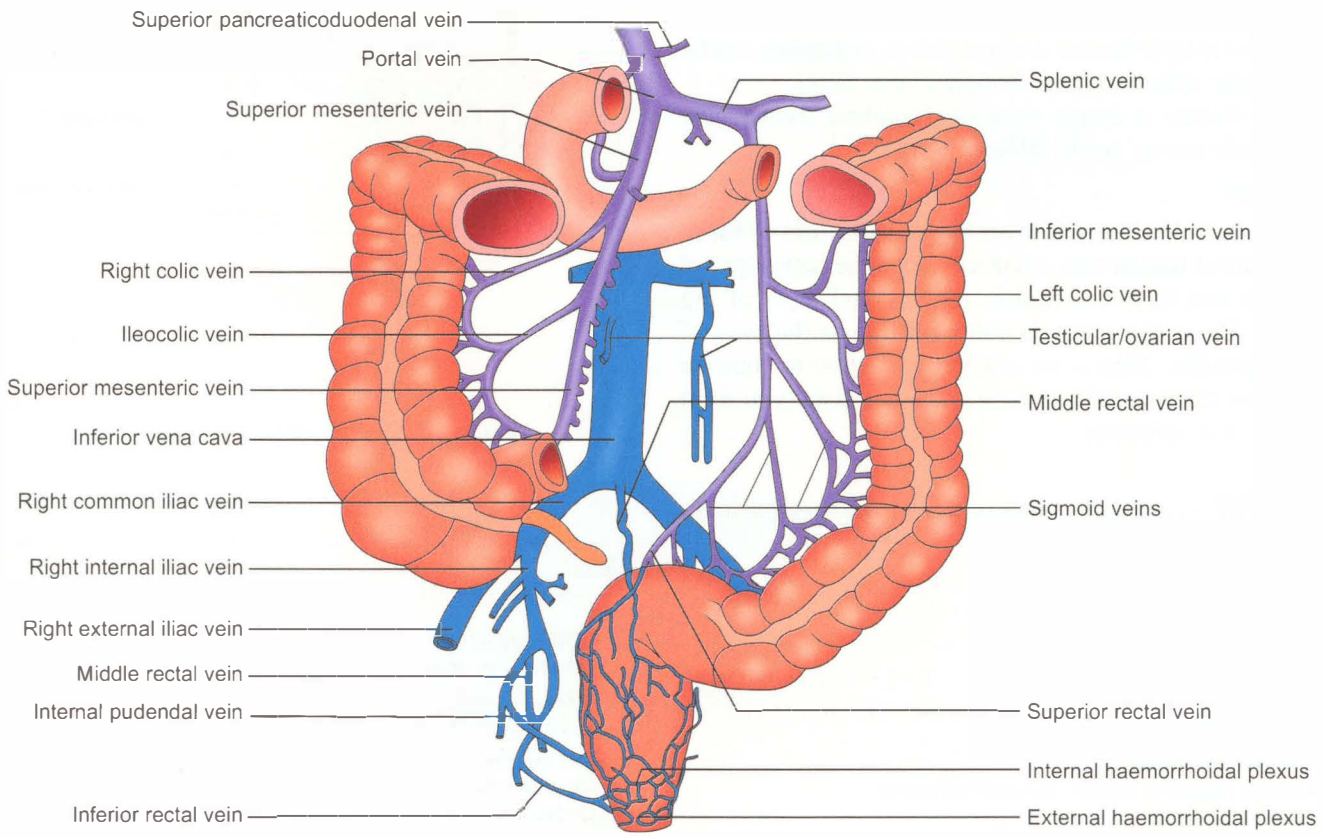


Fig. 29.4: Venous drainage

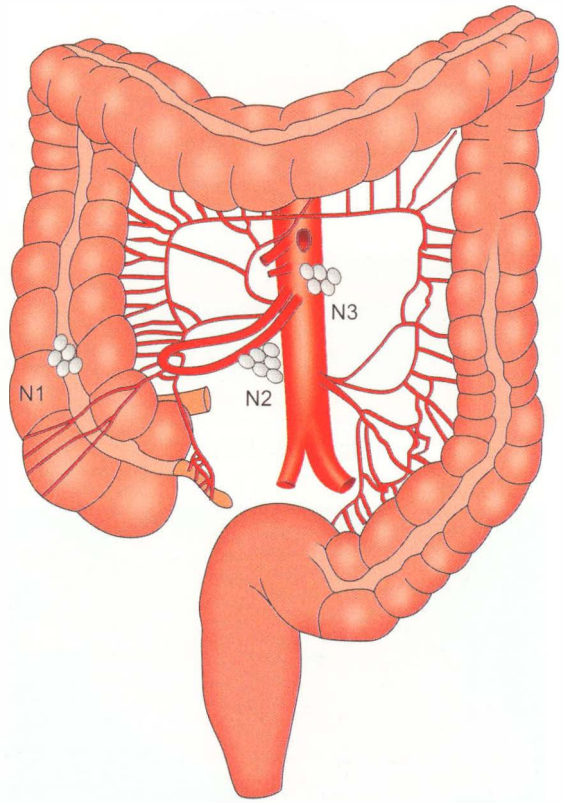


Fig. 29.5: Lymphatic drainage (*see* text for description of N1, N2 and N3)

patients with carcinoma colon at presentation to the hospital. These are called principal nodes.

Colorectal nerve supply

- Sympathetic (inhibitory) arise from T10–T12 and L1–L3
- Parasympathetic (stimulation).
  A. Vagus nerve supplies right and transverse colon.
  B. Sacral nerves (S2–S4 which form nervi erigentes) supply distal colon, i.e. splenic flexure onwards.

Significance

Colonic pseudo-obstruction starts from splenic flexure. Transition zone of vagal supply to sacral nerve supply.

COLONIC FUNCTION

Absorption

Water content of faecal matter is reduced to 1000–1500 ml per day. Thus stools become solid. Similarly, sodium, potassium and bile salts are also absorbed. Significance of this is in cases of diarrhoea, there is loss of water, sodium, potassium and other electrolytes. Amino acids and fatty acids are also absorbed slowly in the colon.

Secretion

Colon also secretes K^+ and Cl^-. It is increased in colitis. Chloride secretion is increased in cystic fibrosis.

Motility

Colon has 4 types of motility. Propulsive, retropulsive, mass peristalsis and gastrocolic reflux. Thus, contents travel aborally. Retropulsive activity is more in the right colon, these allowing the contents to 'churn' more and more. Mass contractions are found more in left colon—specially after meals.

Factors which stimulate the colonic motility

- Dietary fat
- Rich fibre diet
- Physical activity—walking, change in posture, exercises
- Emotional activity
- Less water intake

Constipation (Key Box 29.1)

- It depends upon several factors such as food habits, genetic, social customs.
- Generally a patient is said to have constipation if he passes less than 2 stools per week.
- In addition to the low fibre diet, emotional feelings and many rectal diseases also cause constipation. Example—prolapsed rectum, solitary rectal ulcer syndrome. Colonic disease such as megacolon—Hirschsprung disease is an important cause of constipation in children.
- Increasing constipation in elderly patient suggest carcinoma in left colon. Needs to be evaluated by colonoscopy.

Recycling

- Recycling of various nutrients takes place in the colon. Examples: Fermentation of carbohydrates, short-chain fatty acids and urea cycling.
- Butyrate is the main product of bacterial fermentation. It is required mainly as a fuel for colonic epithelium.
- To accomplish this, the colon depends highly on its bacterial flora, especially for degeneration and fermentation ability.

PEARLS OF WISDOM

More distal the colon—more is the protein metabolism and putrefaction resulting in carcinogens and greater exposure to colonic mucosa. Hence, two-thirds of colonic cancer occur in the left colon.

KEY BOX 29.1

CONSTIPATION

- Digestion and absorption
- In a child—Hirschsprung's disease
- Adult women—idiopathic/following child birth
- Middle aged women—following hysterectomy
- Elderly man—carcinoma left colon
- Constipation with severe pain—anal fissure
- Depression patient—psychotropic drugs—used to treat schizophrenia, antidepressants and antiepileptic drugs.

Colonic bacteria

- Anaerobic bacteria: They constitute more than 99%. The most common pathogen is bacteroides fragilis (10^{10}/g of faeces). Other organisms are clostridia, cocci, etc.
- Aerobic bacteria: *Escherichia coli* is the most common organism about 10^7/g of faeces. Other organisms are Klebsiella, Proteus and Enterobacter.
- Normal function: Bacteria degrade bile pigments thus resulting in brown coloured stools. They also help in colonic motility and absorption. Fatty acids produced by bacteria supply nutrition to colonic epithelium. Bacteria also supply vitamin K to the host.

Prebiotics and probiotics

- **Prebiotics** are non-digestible food ingredients that stimulate the growth and/or activity of bacteria in the digestive system in ways claimed to be beneficial to health. Traditional dietary sources of prebiotics include soybeans, inulin sources (such as Jerusalem artichoke, jicama, soya and chicory root), raw oats, unrefined wheat, unrefined barley, and yacon.
- **Probiotic** is defined as a "live microbial feed supplement which beneficially affects the host animal by improving its **intestinal microbial balance**". They are non-degradable oligosaccharides. They stimulate the growth of beneficial intestinal bacteria.
- Probiotics are dietary supplements which contain live cultures of bacteria and yeast that are beneficial to colonic and host function. The common species used as probiotics are Lactobacillus and Bifidobacterium. Probiotics stimulate immune function, exhibit anti-inflammatory property and suppress pathogenic organism.
- **Clinical application:** When a person takes antibiotics, both the harmful bacteria and the beneficial bacteria are killed. Bacterial change in flora alters carbohydrate metabolism with decreased short-chain fatty acid absorption and result in osmotic diarrhoea. In a similar fashion, antibiotic therapy causes increase in the growth of *Clostridium difficile*. Thus, probiotics have been recommended in antibiotic induced diarrhoea. Also, they have been used to treat diarrhoeas in ulcerative colitis, in pouchitis (inflammation of the pouch after total proctocolectomy for ulcerative colitis) and in necrotising colitis in children.

TUMOURS OF THE LARGE INTESTINE

Benign tumours (Fig. 29.6) are usually referred to as polyp, which means **elevated from the surface**. They are as follows:

ADENOMATOUS POLYP (Key Box 29.2)

- It may be a villous adenoma which is a flat lesion or a tubular adenoma having a pedicle. Tubular is more common.

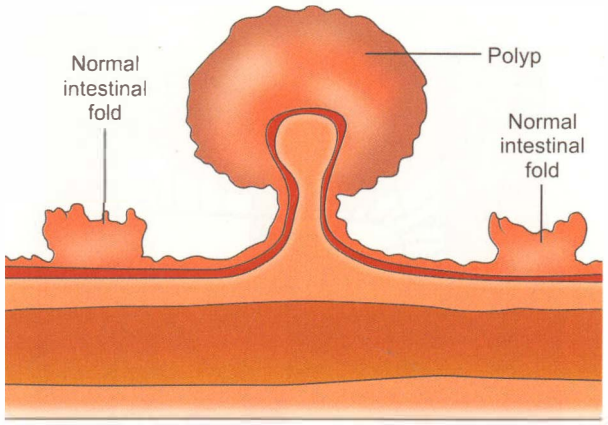


Fig. 29.6: Polyp

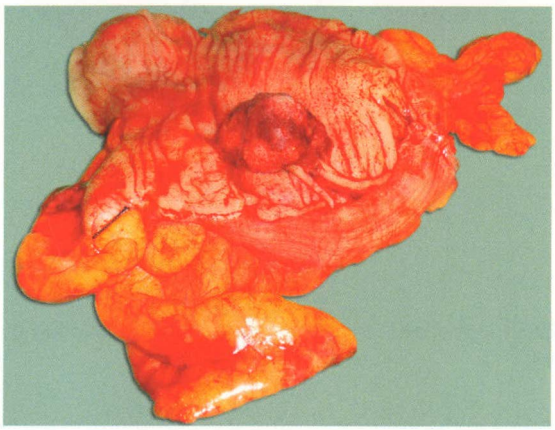


Fig. 29.7: Carcinoma in polyp

- They give rise to bleeding, mucus diarrhoea and hypokalaemia.
- They can be single or multiple
- They are dysplastic
- They are premalignant and the risk of malignancy is greater with increase in the size of the adenoma.
- They can be removed with the colonoscope—polypectomy.
- Malignant potential of ***villous adenoma*** is more than tubular adenoma (Fig. 29.7).
- Adenoma less than 1 cm—risk of malignancy is 1%; 1–2 cm is 10%; > 2 cm is 30%.

KEY BOX 29.2

ADENOMATOUS POLYPS

- Most of the neoplastic polyps occur in elderly patients (> 50 years).
- Most of them are pedunculated
- Most pedunculated polyps are removed by colonoscopic snaring.
- Adenoma larger than 5 mm in diameter carry risk of malignant potential.
- More the polyps, more chances of synchronous carcinoma
- Flat adenomas also carry malignant potential.

- Symptoms and signs of polyps: Bleeding per rectum is the most common symptom. Fresh bleeding is seen in rectal polyps. Typically it is painless. It is intermittent. If it is associated with change of bowel habits means probably a malignant change. These changes include mucus discharge, tenesmus, sometimes constipation. In children, polyp may project outside the anus. In such cases, it has to be distinguished from prolapsed rectum.

Treatment

- Colonoscopy and polypectomy is the standard treatment.
- If specimen shows invasive carcinoma, radical surgery needs should be done.

PEARLS OF WISDOM

Although most neoplastic polyps do not evolve to cancer, most colorectal cancers originate as a polyp.

HAMARTOMATOUS POLYP (JUVENILE POLYP)

- This can occur in the colon as in Peutz-Jeghers syndrome. Risk of malignancy is very limited. Symptomatic polyps need to be treated.
- **Juvenile polyps** are usually single and occur in children. They give rise to bleeding and are easily resected. They do not have malignant potential.

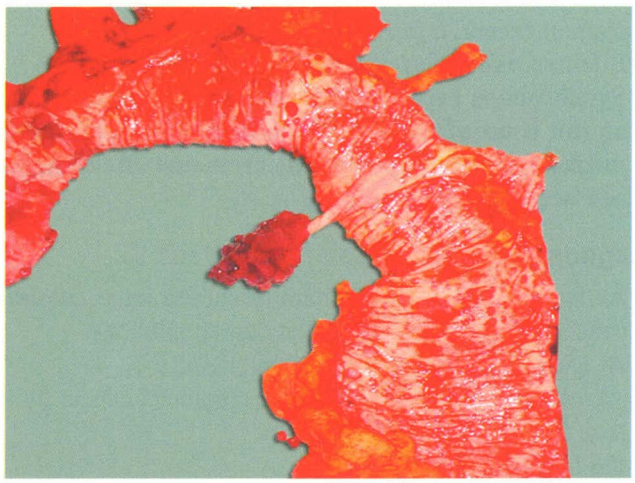


Fig. 29.8: Thousands of polyps. One of them is 2 cm and is pedunculated

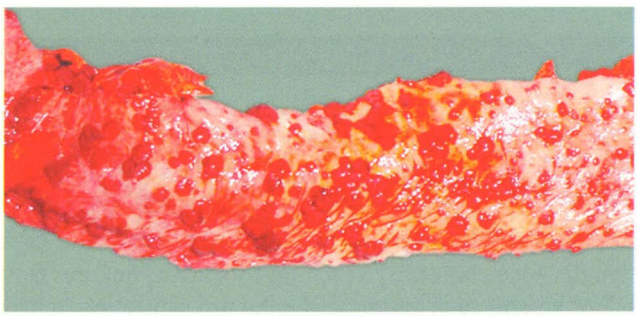


Fig. 29.9: Close-up view of the polyps

FAMILIAL POLYPOSIS COLI (FPC) OR FAMILIAL ADENOMATOUS POLYPOSIS (FAP)

- FAP is a genetic disorder inherited as a Mendelian dominant. The gene APC (adenomatous polyposis coli) is located on the short arm of chromosome 5. Prevalence: 1 in 10,000. It is clinically defined by the presence of more than 100 colorectal adenomas (Figs 29.8 and 29.9).
- It is transmitted from both sexes. The incidence is same in either sex.
- When it is associated with desmoid tumour, craniofacial osteoma, epidermoid cysts, congenital hypertrophy of retinal pigment epithelium, it is described as **Gardner's syndrome** (Key Box 29.3).
- When familial polyposis coli is associated with central nervous system tumour and glioblastoma, it is called **Turcot's syndrome**.
- 50% of them have benign gastric polyps and 90% of them have duodenal polyps.

Clinical features

- Runs in families; other members of the family are affected.
- Manifests at the age of 20 in the form of blood and mucus in the stool, loose stools, etc. It produces crampy lower abdominal pain.
- Anaemia, weight loss and protein malnutrition occur slowly.
- Mean age of development of carcinoma is 39 years.

Complications of FAP

Malignancy (100% risk)

Investigations

Colonoscopy—details in page 707

Treatment

- **NSAID: Sulindac** 300 mg, twice a day and **aspirin** 325 mg once a day have been found to decrease the size of polyps.

KEY BOX 29.3

FAMILIAL POLYPOSIS COLI—SUMMARY

- **P**olyps are more than 100 (colorectal adenomas)
- **O**ther mesodermal tumours—desmoid tumours, osteoma, epidermoid cysts can be present (Gardner's syndrome).
- **L**arge bowel is predominantly involved.
- **Y**ear of development of carcinoma—mean age 39 years.
- **P**olyposis gene—autosomal dominant APC gene.
- **O**ther syndrome—Turcot
- **S**igmoidoscopy from age of 15 at intervals is the investigation of choice.
- **I**leoanal anastomosis with pouch—restorative proctocolectomy—advisable above age of 30.
- **S**urgery is the only means of preventing colonic cancer.

Remember as **POLYPOSIS**

- Patients with FAP who are above the age of 30 have high chances of having a carcinoma in the colon. Hence, even when there is no malignancy, surgery is advisable.

Types of surgery (Figs 29.10 and 29.11)

- Many patients do not like ileostomy. Hence, a subtotal colectomy with ileorectal anastomosis can be done. This is done provided that rectum is examined frequently and endoscopic snaring of the polyps is done regularly, especially in a young patient.
- **Restorative proctocolectomy** with ileoanal anastomosis by using a pouch is another alternative. However, it is a major surgical procedure and should be undertaken only by an experienced surgeon.

Screening

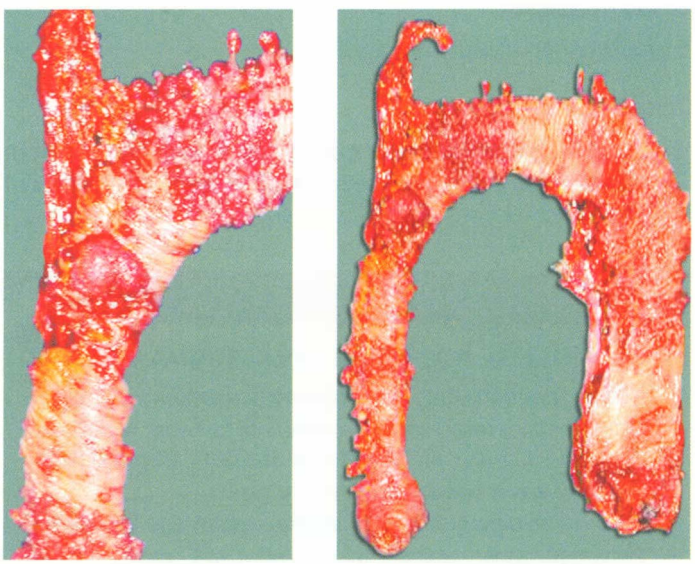
Starts from the age of 15 years using sigmoidoscopy.

PEARLS OF WISDOM

If there are no adenomas by the age of 30 years, FAP is unlikely.

METAPLASTIC POLYP

Also called hyperplastic nodules. They are of viral aetiology. They do not have malignant potential.



Figs 29.10 and 29.11: Familial adenomatous polyposis with two malignancies—lower rectum and hepatic flexure. This patient was being treated for chronic diarrhoea for 7–8 years with various medications. 'He underwent colonoscopy for the first time in our hospital.'Total proctocolectomy specimen (*Courtesy:* Dr Challa Srinivas Rao, Professor, Dept of Surgery, and Dr Ravi, Konaseema Institute of Medical Sciences (KIMS), Amalapuram—Andhra Pradesh)

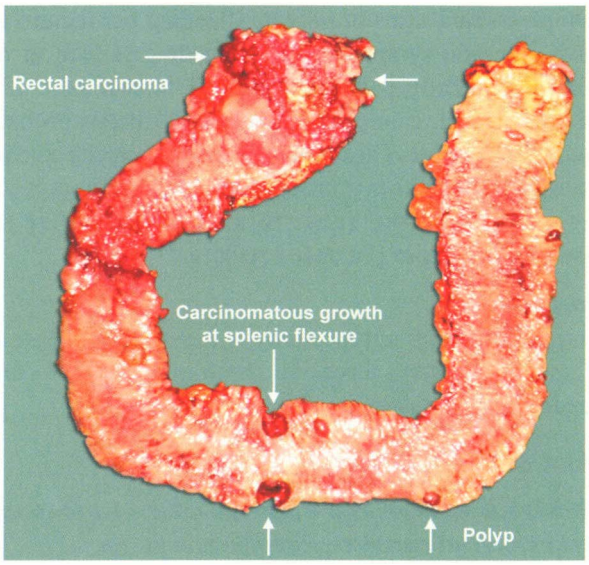


Fig. 29.12: Synchronous carcinoma in adenomatous polyps

HNPCC (HEREDITARY NONPOLYPOSIS COLORECTAL CANCER)

- Autosomal dominant, **no polyps**
- **Lynch's syndrome I:** Site specific colorectal cancer.
- **Lynch's syndrome II:** Cancer family syndrome—they have extracolonic cancers such as endometrial cancer, ovarian cancer, transitional cell cancer, etc.
- Lifetime risk of developing colorectal cancer is 80%.
- **Synchronous carcinoma means more than one cancer at the time of diagnosis. Metachronous carcinoma means appearance of second carcinoma after 6 months can occur here** (Fig. 29.12).

Diagnostic criteria (Amsterdam criteria II)

1. At least 3 members in a family should have colorectal cancer—two of whom are first degree relations.
2. At least two consecutive generations
3. At least one relative should have colorectal cancer by less than 50 years of age.
4. Exclusion of FAP

Screening

Increased incidence of **proximal colonic cancer**.

EXAMINATION OF COLON

Anoproctoscopy: One can examine up to 10–12 cm of anal canal and rectum. Rubber band ligation (for piles) and polypectomy can be done with this instrument.

Flexible sigmoidoscopy: The scope measures about 60 cm in length. One can easily reach up to splenic flexure. Bowel wash or an enema is given before the procedure. No sedation is required (Figs 29.13 and 29.14).

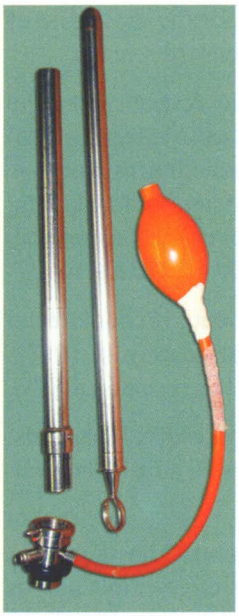


Fig. 29.13: Rigid sigmoidoscope

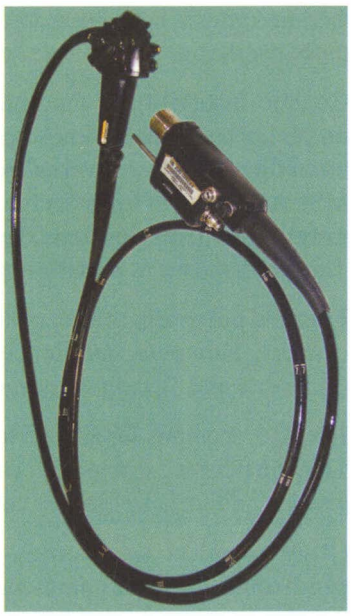


Fig. 29.14: Flexible sigmoidoscope

Fibreoptic colonoscopy can assess the entire colon. It is 100–160 cm in length. Usually there will be multiple polyps varying from a few millimetres to centimetres. Biopsy has to be taken. Polyps are visible after 15 years and certainly by the age of 30 years.

- It is the investigation of choice in most of the large intestinal lesions.
- It permits examination of entire colon and terminal ileum.
- Colon is prepared by polyethylene glycol given orally.
- Risk of perforation of colon is less than 0.1%.

Indications

Diagnostic

- Lower gastrointestinal bleeding
- Inflammatory bowel diseases
- Abnormal finding in barium enema
- Family history of colorectal cancers
- Biopsy of caecum/ileum in suspected cases of cancer.
- Ileocaecal tuberculosis—to take biopsy

Therapeutic

- Control of bleeding—coagulation or injection sclerotherapy.
- Snaring of polyps
- Removal of foreign body
- Detorsion of volvulus
- Decompression of pseudo-obstruction

CARCINOMA COLON

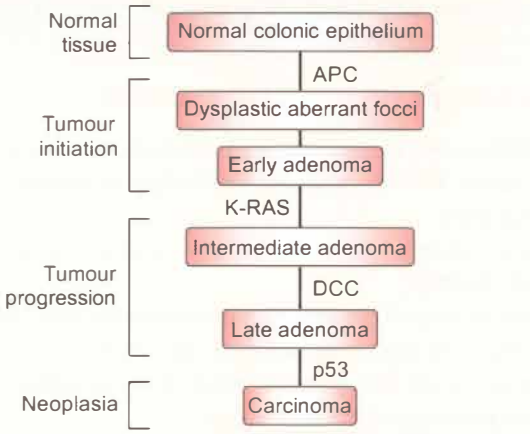
Introduction

It is the second most common cancer and cancer related death cases in the western world next only to lung cancer. The incidence increases with age. Multiple synchronous lesions (more than 1 malignancy at the time of diagnosis) is found in about 5% of the patients. Colon is also one of the sites of metachronous cancer (new malignancy appearing after 6 months of curative surgery). More than 95% are adenocarcinoma and surgery remains the most effective treatment. Survival has improved because of early diagnosis and multimodality of the treatment.

Over a period of years the understanding of development of carcinoma has changed and more and more molecular biology of colonic cancer is being discussed. The **Fearon Vogelstein adenoma-carcinoma multistep model** of colorectal neoplasia represents one of the best-known models of carcinogenesis (Key Box 29.4).

KEY BOX 29.4

FEARON-VOGELSTEIN MODEL



APC, K-RAS, DCC and p53 are oncogenes that convert normal cell to cancerous cell

- **Terminology:** Before we start the discussion on carcinoma colon, we shall study a few terminologies used in carcinoma colon. They are synchronous carcinoma, metachronous carcinoma, familial colorectal carcinoma.

SYNCHRONOUS CARCINOMA

- Moertel's definition: Synchronous cancers as those occurring within 6 months of the first primary cancer, or two or more histologically distinct simultaneously detected malignancies or more than one malignancy at the time of initial diagnosis. This will happen especially in cases of colon and upper aerodigestive tract wherein the stimulus or aetiological factor for malignancy affects different parts of the organ.
- Colon, head and neck, oesophagus are the site of synchronous carcinomas.
- In cases of carcinoma colon with specific aetiological factors such as familial polyposis coli, ulcerative colitis, hereditary non-polyposis cancer, often carcinoma is synchronous.
- Thus, it is important to do a complete colonoscopy when a patient comes with colonic carcinoma because he/she may be having another synchronous carcinoma elsewhere. *See* the case report in the next page.

CLINICAL NOTES

A 68-year-old lady was admitted with large bowel obstruction. Plain X-ray abdomen showed intestinal obstruction. Exploratory laparotomy was done. A 3 cm constricting growth was identified at rectosigmoid junction and high anterior resection and anastomosis was done. On the 4th postoperative day, the patient was allowed liquid diet. Distension increased. For another 3 days, distension went on increasing. Plain X-ray abdomen revealed obstruction with more gas than before. The patient was having colicky abdominal pain. Exploratory laparotomy was done. Findings at 2nd laparotomy—anastomosis was intact. Transverse colon was hugely dilated. Careful palpation of splenic flexure revealed one more growth. A resection and anastomosis was done again. Patient was discharged after 10 days. The first surgeon agreed that after finding out the rectosigmoid growth, he did not look for any other lesions (mistake). This was obviously a case of synchronous carcinoma.

METACHRONOUS COLONIC CANCER

- Metachronous cancer was defined as those cancer occurring more than 6 months following resection of one malignancy.
- A few examples of metachronous site are: Colorectum, breast, kidney.
- Family history of hereditary, nonpolyposis colorectal cancer (HNPCC or Lynch syndrome), an autosomal dominant disease, also can present with both synchronous or metachronous colorectal cancers.
- It is more common in females
- Common usually at young age.
- The associated genetic defect lies at the mismatch repair genes, responsible for the correction of DNA bases mismatch.
- The coexistence of adenomatous polyps is also considered a risk factor for the development of metachronous lesions. Thus, after treating one carcinoma, example—carcinoma sigmoid, annual colonoscopy is recommended. If polyps are detected, patients have to be informed about the polyps and their potential of malignancy.
- Survival is better

FAMILIAL COLORECTAL CANCER

- All these have a carrier gene and thus, run in families. Malignancies occur in young age group. Often they are synchronous. Metachronous lesions are not uncommon.
- Certain criteria have been laid upon for the diagnosis of these conditions which have been discussed already.
- Genetic instability is the chief factor responsible. The instability can be at chromosomal level called chromosomal instability or at DNA level called microsatellite instability—MSI. As a result of this after the cell division by duplication, mismatched genes develop. These genes cannot be repaired. This predisposes to mutation, results in a cancer gene.
- **Familial polyposis coli** accounts for about 1% of colorectal cancers. However, incidence of malignancy is 100%. Gardner's and Turcot's syndrome are the variants of FPC.
- **HNPCC** accounts for about 5 to 10% of colorectal cancers. They also have extracolonic cancers such as endometrial, ovarian and urinary bladder cancers.
- Other familial syndromes are **Cronkhite Canada syndrome.** It is more common in females. Multiple polyps develop in stomach, duodenum and in the colon. Diarrhoea is the clinical presentation. Other features include pigmentation, alopecia, loss of weight and cachexia. Chances of developing malignancy is about 15%.

Precancerous conditions (Key Box 29.5)

1. **Polyps:** Environmental and genetic factors favour the development of colonic polyps and their transformation into malignancy. The incidence of malignancy is increased when the polyp is more than 1 cm, polyps are multiple or flat (Table 29.2).

KEY BOX 29.5

RISK FACTORS ASSOCIATED WITH COLON CANCER

Risk factor	Incidence of cancer
Familial polyposis coli	100% chances of colorectal cancer
HNPCC	80% chances
Ulcerative colitis	10–20% after 20 years
Crohn's colitis	5% after 10–20 years
Adenomatous polyps	1–20% depending upon the size

Table 29.2 Summary of colonic polyps

Type	Cause	Malignant potential	Features/syndrome
1. Adenomatous polyp	Benign tumour	1–10%	Hypokalaemia, diarrhoea
2. Hamartomatous polyp	"Misfire"	Negligible	Peutz-Jeghers syndrome
3. Familial polyposis coli	Genetic disorder	100%	Gardner's and Turcot's syndrome
4. Metaplastic polyps	Hyperplasia	Nil	Asymptomatic

PEARLS OF WISDOM

Familial polyposis coli has 100% chance of carcinoma.

2. Inflammatory bowel disease

A. Ulcerative colitis is a definite precancerous condition. Presence of dysplasia diagnosed by colonoscopic biopsy is an indication for colectomy.

B. Crohn's involving colon also has a mildly increased risk of developing carcinoma when compared to ulcerative colitis.

C. Schistosomal colitis: The risk of colorectal cancer is increased in patients with long-standing schistosomal colitis. Long-standing cases are associated with mild to severe grades of colonic epithelial dysplasia. Thus ulcers or pseudo-polyps can occur. These dysplastic changes are considered as premalignant.

D. Radiation exposure: Usually it is mucin secreting adenocarcinoma with poor prognosis.

E. Ureterosigmoidostomy increases risk of colonic cancer over 100–500 times.

Aetiological factors

1. **SAD factors:** It is sad to know that **SAD** factors are responsible for carcinoma colon. They are **S**—Smoking, **A**—Alcohol, **D**—Dietary factors. Diet rich in red meat has high animal fat. This alters intestinal bacteria, which convert primary bile acids into secondary bile acids. ***This is the beginning of formation of carcinogenic polycyclic aromatic compounds.*** After cholecystectomy, there is increase in free bile acid concentration, thus increasing the risk of colonic cancer. Thus increased roughage is associated with increased transit time which in turn reduces exposure of mucosa to carcinogens.

- Increase incidence is found in western countries wherein diet rich in animal fat is consumed in large quantity. Incidence is more after the age of 50 years. Obesity, poor exercise and smoking are the contributing factors.
- Some interesting observations are also found in females with colonic cancer which have been depicted in Key Box 29.6.

KEY BOX 29.6

WOMEN AND COLONIC CANCER

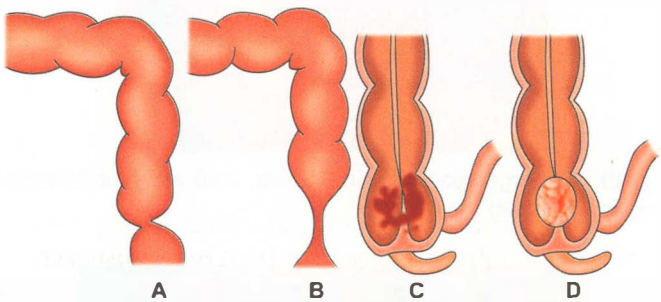
- **Caecum** is more commonly involved than other parts.
- Women with **breast cancer** have increased incidence of colonic cancer.
- Women **smokers** have increased chances of colonic cancer.
- Women who have undergone **cholecystectomy** have increased chances of colonic cancer.
- Metastasis to ovary is mostly haematogenous (1 to 10%).

PEARLS OF WISDOM

Calcium salts are protective. They form insoluble bile salt complexes, thus reducing the concentration of bile acids in the colon.

Pathological types (Fig. 29.15)

- It is an adenocarcinoma—columnar.
  Rectum (40%) and sigmoid (20%) take a major share in colorectal carcinoma followed by caecum (12 to 15%). Multiple synchronous cancers are also common in the colon.
  A. Annular stricture: Common in left colon (splenic flexure, pelvic colon).
  B. Tubular stricture: Common in left colon and at the rectosigmoid junction.
  C. Ulcerative lesion: Ascending colon or caecum.
  D. Proliferative growth: More in right colon, the least malignant, fleshy and bulky polypoid lesion (Fig. 29.16).
- It is a columnar cell adenocarcinoma. In about 5% of cases, it undergoes mucoid degeneration. Such tumours carry poor prognosis. They spread to the liver very fast and secondaries produce mucoid material.



Figs 29.15A to D: Types of carcinoma colon

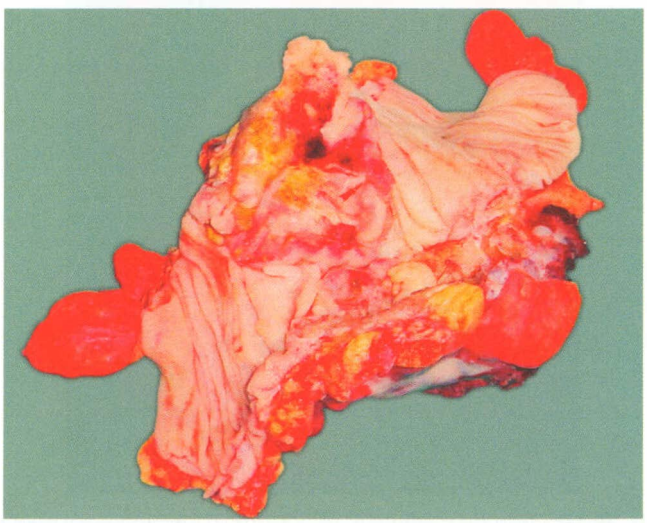
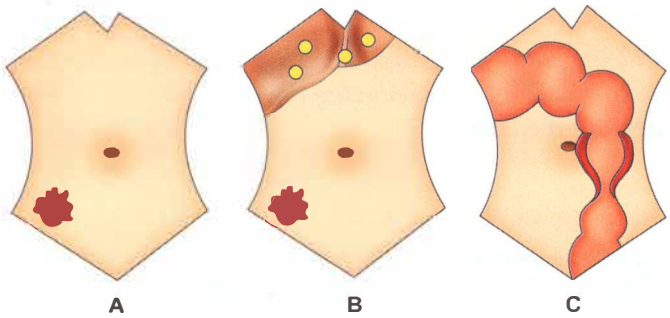


Fig. 29.16: Ulceroproliferative growth in the descending colon



Figs 29.17A to C: (A) Carcinoma caecum, (B) Carcinoma caecum with secondaries in the liver, (C) Carcinoma left colon with intestinal obstruction

Fig. 29.18: Large secondary in the liver. Look at the umbilication (central necrosis)

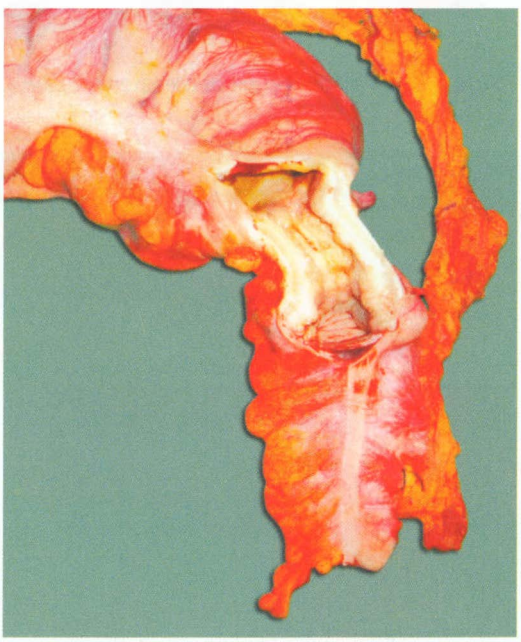


Fig. 29.19: Tubular stricture at rectosigmoid junction—cut opened specimen after resection

Clinical features of carcinoma colon (Figs 29.17A to C and Table 29.3) (Mnemonic TMA Pai)[1]

1. **Tumour:** The mass produced by carcinoma caecum and even hepatic flexure is palpable. It is firm to hard, irregular and with or without fixity.
   - Occasionally, growth at pelvirectal junction can be felt on rectal examination.
   - However, on left-sided constrictive lesions, growth is not often felt. It is the hard faecal matter and lymph nodes which are felt as a mass.
2. **Metastasis:** 5–10% of the patients present with metastasis to liver (mucoid adenocarcinoma), ascites, etc. Distant metastasis is not common (Fig. 29.18).
3. **Anaemia** is an important feature of carcinoma caecum. It may be due to blood loss or a proliferative growth secreting toxins causing suppression of bone marow. Asthaenia and anorexia are the other features.
4. **Pain abdomen:** Dull aching pain may be present. Colicky pain is due to chronic obstruction as in left-sided growths (napkin ring stricture).
5. **Alteration in the bowel habits:** A recent constipation, increase in the dose of laxatives followed by attacks of diarrhoea can be due to carcinoma colon. Diarrhoea is due to hard faecal balls, irritating the colonic mucosa resulting in increased secretion of mucus produced by proximal colon.
6. **Intestinal obstruction** is caused by constricted left-sided lesions (Fig. 29.19). On the left side, diameter of the colon is narrow, contents are solid and growth is constrictive. Lower abdominal distension, right to left peristalsis are the

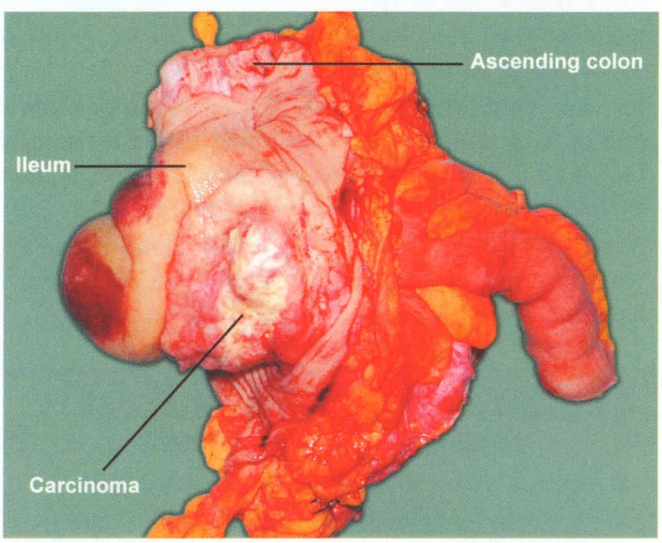
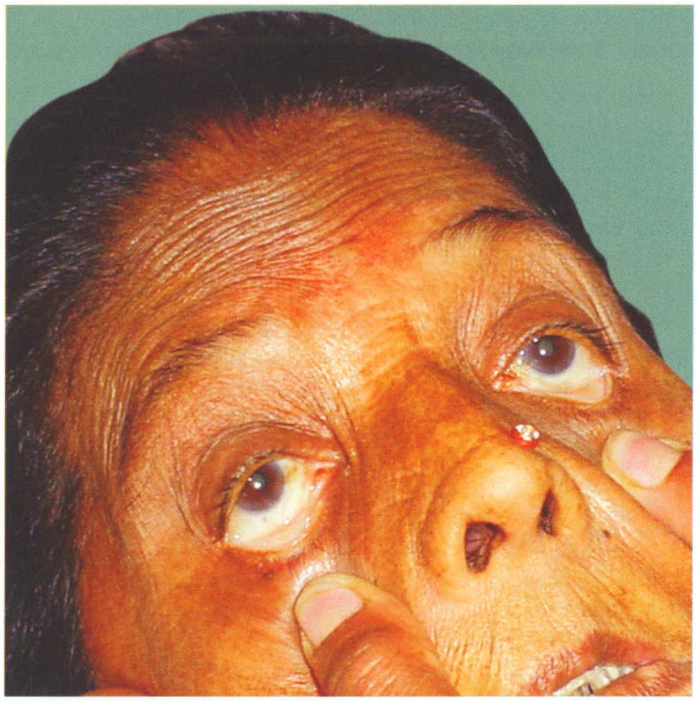


Fig. 29.20: Intussusception due to carcinoma caecum

[1]Late Padmashree Dr TMA Pai, was the founder of Kasturba Medical College which he started in 1953. He was also a banker, educationist. He is called 'Modern Architect of Manipal', where 'Manipal University' is situated.



29.21: Pallor disproportionate to the blood loss—typical of carcinoma caecum

late features. Carcinoma sigmoid can cause colovesical fistula.

To summarise

- **Early cases:** It can be easily missed—such as change in bowel habits like diarrhoea, vague ill health, weakness (due to anaemia), intermittent bleeding per rectum, often attributed to piles or some other cause. Mass is usually not palpable.
- **Late cases:** It can present with obstruction (rectosigmoid junction growth), perforation, intussusception (right-sided tumours), mass abdomen, secondaries in the liver, left supraclavicular nodes (Troisier's sign), etc.
  See Key Boxes 29.7 to 29.9.

KEY BOX 29.7

PECULIARITIES OF CARCINOMA OF CAECUM (Figs 29.20 to 29.26)

- Incidence is more in females
- Presents with anaemia and a mass
- Can present as acute appendicitis when the lumen of appendix is obstructed (in elderly patients).
- It is a cause of intussusception (secondary)
- Can present with flexion of hip due to infiltration of iliopsoas.

KEY BOX 29.8

PECULIARITIES OF CARCINOMA SPLENIC FLEXURE

- Presents as obstruction
- It is easily missed unless carefully looked for
- Carries poor prognosis as many cases present with obstruction.
- Often, it is inoperable

KEY BOX 29.9

PECULIARITIES OF CARCINOMA SIGMOID (Figs 29.27 and 29.28)

- Commonly presents as **obstructive lesion**—constipation and intestinal obstruction.
- **Colovesical fistula:** Carcinoma sigmoid is the 2nd commonest cause.
- It can cause **colovaginal fistula**
- It can also **infiltrate ureter, uterus and ovary.**
- It can present as **abscess in the lateral abdominal wall.**

PEARLS OF WISDOM

Obstructed and perforated carcinoma colon have poor prognosis.

Spread

1. **Local:** For a long time, the lesion is confined to mucosa and submucosa. They grow in annular fashion and later

Table 29.3 Carcinoma right colon and carcinoma left colon

	Carcinoma right colon	Carcinoma left colon
• Presentation	Unexplained weakness, anaemia	Change in bowel habits
• Bleeding	Occult blood in stools	Gross blood in stools
• Abdominal discomfort	Right side and dyspeptic symptoms also	Constipation and obstruction
• Incidence	More common in women	More common in men
• Frequency	About 10–20%	About 60–70% (including rectum)
• Pathology	Ulcerative/proliferative lesion	Strictures (rectosigmoid)
• Investigation	Colonoscopy	Flexible sigmoidoscopy
• Complication	Obstruction—less common	Obstruction, perforation, pericolic abscess—common

longitudinally. Once serosa is involved, spread occurs rapidly into neighbouring structures such as ureter, bladder, uterus, etc. The involvement of these structures is **not a contraindication** for surgery (TNM staging).

- Local perforation may result in pericolic abscess.
- Hollow viscus perforation results in internal fistula.

2. **Lymphatic spread** (*see* page 702)
3. **Blood spread:** It occurs late, resulting in secondaries in the lungs, liver, etc. Cannon ball in lung nodule in the liver.

PEARLS OF WISDOM

Because of the drainage into the portal system, colonic cancers spread to the liver first. On the other hand, rectal cancers spread to the lungs because of drainage into inferior vena cava.

STAGING/CLASSIFICATIONS

There are many classifications and staging for carcinoma colon/rectum. They are not important. A few important ones have been given here.

I. Dukes' staging for colorectal cancer

- **Stage A:** Invasion of but not breaching the muscularis propria.
- **Stage B:** Breaching the muscularis propria but not involving the lymph nodes.
- **Stage C:** Lymph nodes are involved

  Few authors describe a stage D for metastatic disease. Since it has not been described by Duke, it is called modified Dukes' staging.

II : Astler-Coller's modification of Dukes' staging

Stages (Fig. 29.29)

A	Limited to mucosa—no nodes
B1	Extension into muscularis propria—no nodes
B2	Extension into entire bowel wall—no nodes
B3	Extension into adjacent organs—no nodes
C1	Extension into muscularis propria—positive nodes
C2	B2 + Lymph nodes
C3	B3 + Lymph nodes
D	Distant metastasis

III. WHO classification—it is based on histology

- Majority are adenocarcinoma—90%
- Mucinous adenocarcinoma—5–10%
- Signet ring cell carcinoma
- Small cell carcinoma
- Squamous cell carcinoma
- Undifferentiated carcinoma

IV. TNM staging (*see* text on the right side)

A few clinical photograph, staging pictures and operative pictures have been given in next page.

TNM STAGING OF COLORECTAL CANCER

Tumour—T

Tx—Primary tumour cannot be assessed
T0—No evidence of tumour
Tis—Carcioma *in situ*—intraepithelial/invasion into lamina propria
T1—Invasion into submucosa
T2—Invasion into muscularis propria
T3—Invasion into pericolorectal tissues/fat
T4a—Invasion into surface of the visceral peritoneum
T4b—Direct invasion or adherent to adjacent structures/organs

Regional nodes—N

Nx—Nodes cannot be assessed
N0—No nodal spread
N1—Regional nodes 1–3 involved
– N1a—1 regional node
– N1b—2 to 3 regional nodes
– N1c—Tumour deposits in serosa/mesentery/non-peritonealised pericolic or perirectal tissue without regional nodes
N2a—Regional nodes 4 or more involved
– N2a—4–6 regional nodes
– N2b—7 or more regional nodes

Distant metastases M

M0	No distant spread
M1	Distant spread present
M1a	Spread confined to one organ or site—liver/lung/ovary/ nonregional nodes
M1b	Spread to more than one organ or site/peritoneum

Histological grade G

Gx—Grade cannot be assessed
G1—Well-differentiated
G2—Moderately differentiated
G3—Poorly differentiated
G4—Undifferentiated

Residual tumour R

R0—No residual tumour after resection
R1—Microscopic residual tumour after resection
R2—Macroscopic residual tumour after resection

Stage Group

0—Tis N0 M0
1—T1 N0 M0; T2 N0 M0
IIA—T3 N0 M0
IIB—T4a N0 M0
IIC—T4b N0 M0

IIIA—T1-2 N1-1c M0; T1 N2a M0
IIIB—T3-4a N1-1c M0; T2-3 N2a M0; T1-2 N2b M0
IIIC—T4a N2a M0;T3-4a N2b M0;T4b N1-2 M0
IVA—Any T any N M1a
IVB—Any T any N M1b
V0—No venous invasion;V1—presence of venous invasion
L0—No lymphatic vessel invasion; L1—presence of lymphatic vessel invasion

Fig. 29.22

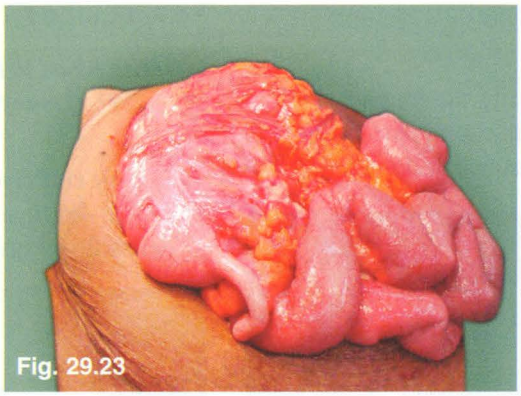


Fig. 29.23

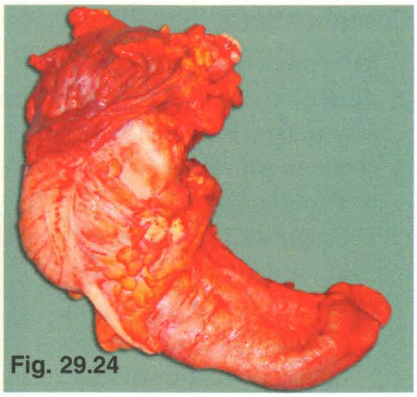


Fig. 29.24

Fig. 29.22: A 34-year-old lady presented to the hospital with mass in the right iliac fossa with slight flexion of the right hip. Mass was hard and irregular. She also had anaemia **Fig. 29.23:** Exploration of the mass (caecum). It was mobile **Fig. 29.24:** Right hemicolectomy specimen—a case of carcinoma caecum

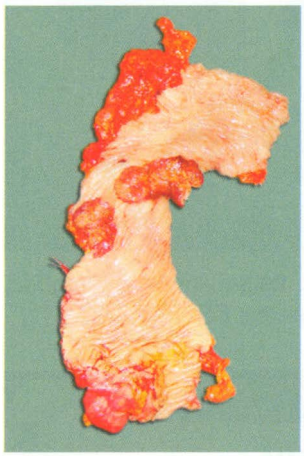


Fig. 29.25: Carcinoma hepatic flexure with partial obstruction

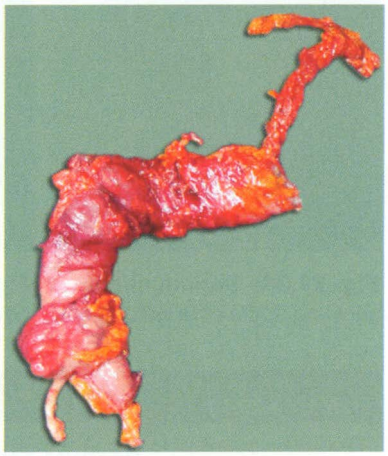


Fig. 29.26: Extended right hemicolectomy specimen

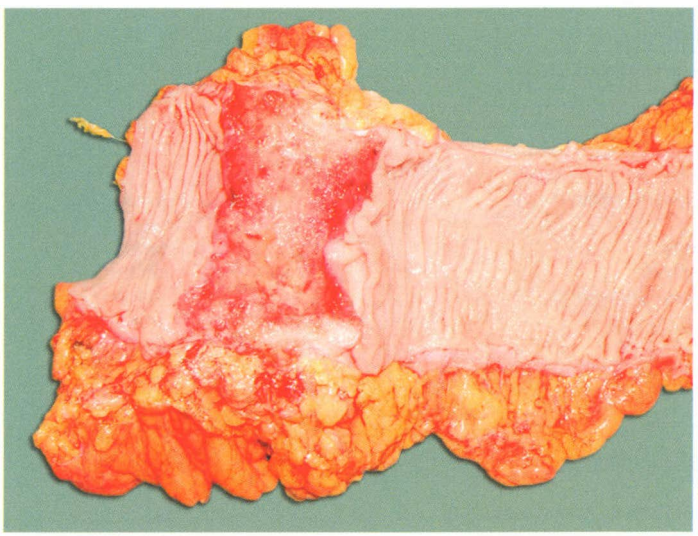


Fig. 29.27: Rectosigmoid stricture. Patient underwent high anterior resection. 5 cm proximal margin is enough for radical cure

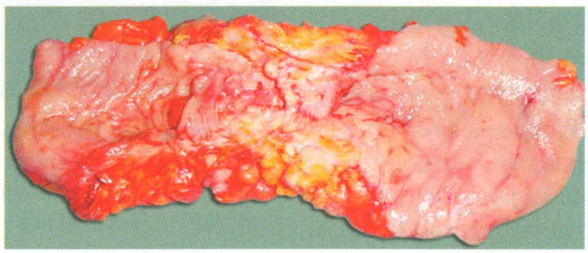


Fig. 29.28: Intestinal obstruction due to rectosigmoid stricture—a common complication. **Tumour rarely goes beyond 2 cm from the edge of the tumour** unless there is concomitant spread to lymph nodes

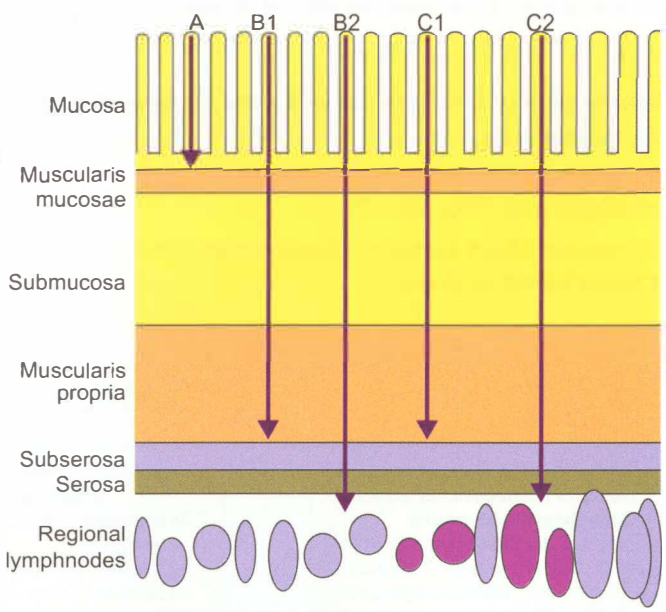


Fig. 29.29: Astler-Coller staging

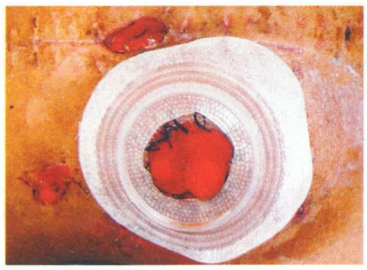


Fig. 29.30: Limited colectomy leak—managed by refashioning of the stoma followed by ileostomy

Complications (Fig. 29.30)

1. **Intestinal obstruction**
2. **Pericolic abscess:** Pain is present in the tumour site and may radiate to back, leg or hip as in caecal perforations. It is due to irritation of the psoas muscles or due to irritation of femoral nerve.
   - Diagnosis is confirmed by ultrasound/CT scan.
   - Percutaneous aspiration, followed by elective resection is the best treatment.
3. **Faecal fistula** (Fig. 29.31)
   - Pericolic abscess when it is incised or drained to the exterior may result in faecal fistula if there is malignancy.
   - Carcinoma caecum may result in appendicitis and appendicectomy may invariably result in faecal fistula.

Fig. 29.31: Carcinoma colon perforated—pericolic abscess which on draining resulted in faecal fistula

4. **Internal fistula:** Colovesical, colocolic, coloenteric are not uncommon complications of malignancies. They are managed by resection. However, preoperative assessment of fistulae by investigations should be done.

PEARLS OF WISDOM

Involvement of local structures is not a contraindication for radical resection.

Investigations (Fig. 29.32)

1. **Complete blood picture**—demonstrates low Hb%
2. **Occult blood** in stools

PEARLS OF WISDOM

Occult blood in the stools may be the finding which gives a 'clue' in many cases of 'anaemia for evaluation'.

3. **Double contrast barium enema** may show irregular filling defect—intrinsic, persistent. It may also show an apple core deformity (Figs 29.33 and 29.34 and Key Box 29.10).

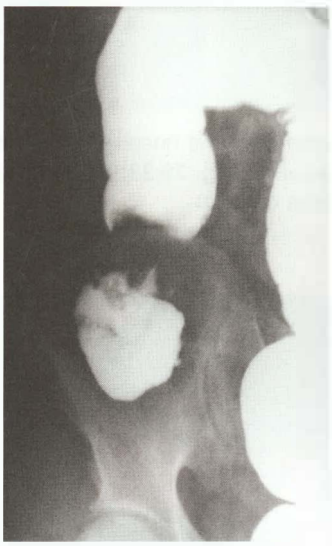


Fig. 29.33: Barium enema showing apple core deformity

Fig. 29.34: Intrinsic, irregular, persistent filling defect

KEY BOX 29.10

BARIUM ENEMA

- Gives good anatomic and topographic information.
- Can detect associated diverticular disease.
- Small ulcerative lesions can also be diagnosed.

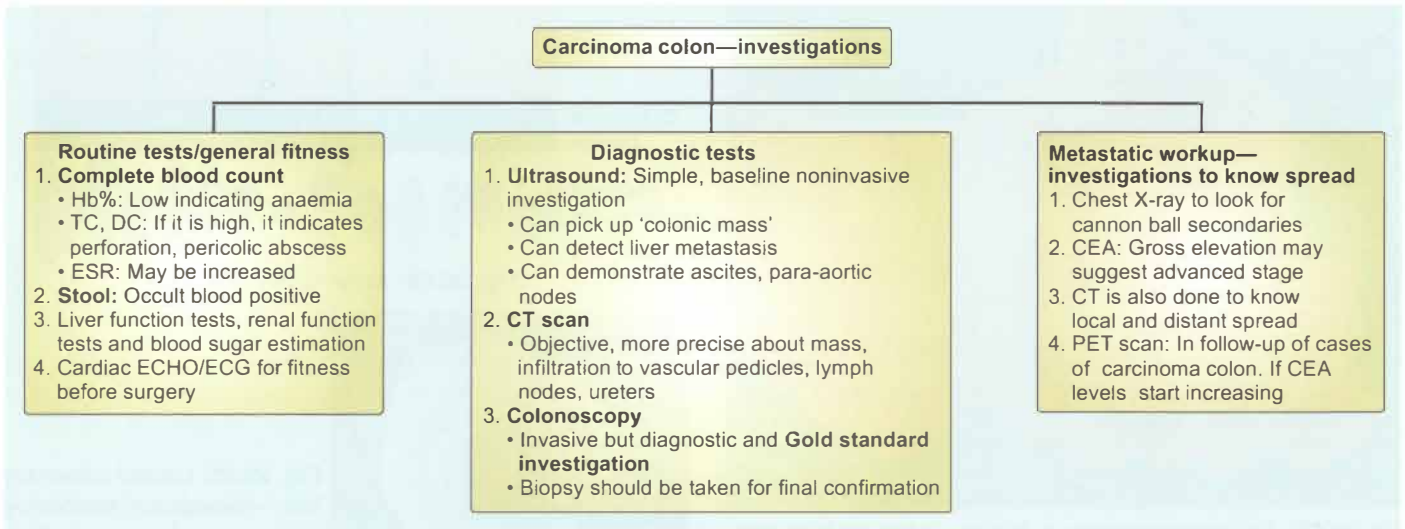
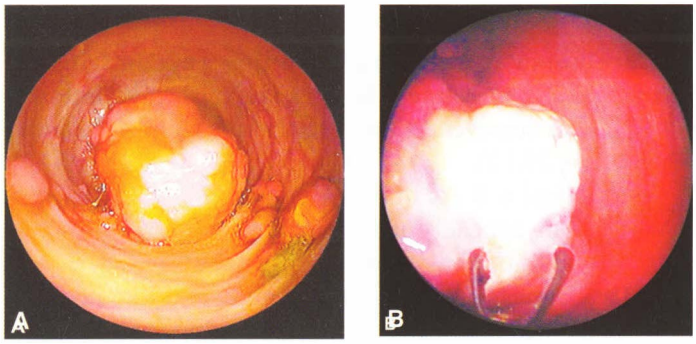


Fig. 29.32: Carcinoma colon—investigation



Figs 29.35A and B: Colonoscopy shows growth and biopsy is being taken (*Courtesy:* Dr Filipe Alvares, Consultant, Medical Gastroenterologist, KMC, Manipal)

4. **Flexible sigmoidoscopy:** 60 cm of colon can be visualised. It is an outpatient procedure. It is indicated in rectal bleeding. An enema is given before the procedure.
5. **Colonoscopy** is done to take a biopsy from growth and also to rule out synchronous malignancy as seen in 5% of the cases (more than one malignancy at the time of diagnosis). If biopsy cannot be taken, as in obstruction, brush cytology can be taken (Fig. 29.35).
   - Small risk of perforation is present and it is invasive procedure.
   - **Virtual colonoscopy** can pick up polyp of 6 mm size also but biopsy cannot be taken.
6. **Ultrasound:** It is the baseline investigation to be done first.
   - It can detect colonic mass
   - It can detect hydronephrosis, liver metastasis, ascites, para-aortic nodes.
   - Ultrasound guided biopsy is possible in advanced cases.
   - Definitely it cannot pick up early mucosal or lesions which have penetrated up to muscularis mucosa or serosa.
7. **CT scan in carcinoma colon**
   - Other than a biopsy, it has all the advantages and it is the investigation of choice after colonoscopy.
   - Anatomical location of the tumour, involvement of serosa
   - Infiltration of local adjacent structures like ureter (right side)—preoperative stenting is required in such cases.
   - Metastasis in the liver, ascites, para-aortic nodes.
   - Other associated diseases specially in elderly such as aortic aneurysm, gall stones, hiatus hernia, etc.
   - 90% and 95% sensitivity and specificity in detecting liver lesions greater than 1 cm.
8. **Carcino-embryonic antigen (CEA)** (Key Box 29.11).
   - It is a foetal glycoprotein, not present in normal human beings (minute quantities). It is present in the cell membranes of many tissues including colorectal cancer.
   - It is present in the last trimester in the foetus.
   - It has a prognostic rather than a diagnostic value. After treatment of the primary, CEA level should come back to normal. If it is increased, it suggests either recurrent tumour or secondaries in the liver.

KEY BOX 29.11

CARCINO-EMBRYONIC ANTIGEN—CEA

- Discovered by Gold and Freedman
- It is a surface glycoprotein
- Produced by colorectal epithelium but cleared by Kupffer cells of liver. Its half-life is prolonged (normal—10 days) in cholestasis and hepatocellular dysfunction.
- Normal levels: 0–4 mg/ml
- Significant increase in the levels is also found in pancreatic carcinoma, gastric carcinoma, lung carcinoma, breast carcinoma.
- Increased CEA in the follow-up period of colorectal cancer suggests metastasis. PET scan may help in these patients when CT/US are normal.
- It has very low sensitivity
- If preoperative CEA is increased in node-negative colonic cancer, chemotherapy is recommended.

9. **Role of PET scan**
   - Routine use of PET—positron emission tomographic scanning in the primary management of colorectal cancer is not recommended.
   - It is useful in the follow-up cases wherein CEA levels are increasing and the actual cause for the rise is being evaluated.
   - **Chest X-ray:** Colonic carcinoma spreads more often to lungs than carcinoma of stomach giving rise to cannon ball secondaries.

Prognostic factors of carcinoma colon

- **Spread:** If it is limited to mucosa and there are no nodes, 5-year survival is 90–100%.
- **Age:** Younger patients have poor prognosis.
- **Grade:** Poorly differentiated tumours have worse prognosis.
- **Obstruction and perforation:** Poor prognosis due to dissemination of malignant cells.
- **Blood transfusion:** Perioperative blood transfusion has poor prognosis.

PEARLS OF WISDOM

Blood transfusion increases number of suppressor T-lymphocytes thus causing immunosuppression. Hence, follow bloodless surgery or autologous blood transfusion.

Preoperative preparation

1. **Mechanical bowel preparation** is necessary to reduce the bacterial load in the colon. This reduces incidence of anastomotic leakage.
   - **Whole gut irrigation by oral polyethylene glycol** is found to be superior than enemas. It is the method of choice today. It is mixed with 2 litres of water and is given 12 hours before surgery.

2. **Antibiotics:** Oral antibiotics neomycin/metronidazole or neomycin/erythromycin are given in the afternoon and evening before surgery. They act locally and are given mainly to decrease wound infection.

   Intravenous antibiotics such as ciprofloxacin and metronidazole or 3rd generation cephalosporins such as ceftriaxone is given one hour before the surgery. The target bacteria are *E. coli* and *Bacillus fragilis*.
3. Improvement of **general condition** by correcting albumin levels and if necessary, TPN will definitely decrease the incidence of anastomotic leakage.
4. A **fat-free diet, low residue diet** is prescribed two to three days before surgery.
5. **Prophylactic fractionated heparin** enoxaparin or dalteparin is given subcutaneously, to prevent deep vein thrombosis. Otherwise subcutaneous heparin is given.

Principles of surgery

Colonic surgery is a very common problem encountered in the practice or institution. Every surgeon should be aware of basic principles of colonic surgery and technique of anastomosis. I have grouped them together under the heading of ten commandments so as to understand better.

Surgery

Over a period of years, radical resections for carcinoma of colon have become less radical pertaining to the extent of bowel resection. For example: One need not remove terminal 30 cm of ileum for carcinoma caecum today. Only 6–8 cm of ileum removal is sufficient in a right hemicolectomy.

TEN COMMANDMENTS OF SURGERY FOR CARCINOMA COLON—OPEN SURGERY

1. Should mark the ostomy site preoperatively—in cases of emergency colectomy.
2. Should give an adequate incision.
3. Should explore the peritoneal cavity for metastasis.
4. Should remove the growth with at least 7 cm margin, with all groups of regional nodes, fat fascia and lymphatics called *en block* resection—R-0 resection.
5. Should do the resection without touching or handling the tumour—follow no touch technique of Turnbull.
6. Should divide the vascular pedicle first and should ligate the vessels as high at the origin—High tie.
7. Should ensure the cut ends bleed well before anastomosis.
8. Should ensure there is no tension at the suture line.
9. Should do one stage procedure in all elective cases—resection and anastomosis.
10. Should consider temporary ileostomy after resection anastomosis in obstructed colon cancer.

Different types of surgery

1. **Carcinoma right colon including caecum:** If it is operable, the treatment is right radical hemicolectomy. Structures removed in this operation are (Fig. 29.36 and Key Box 29.12).
   - **Terminal 6–8 cm of ileum**[1]
   - Caecum, appendix and ascending colon
   - One-third of transverse colon
   - Fat, fascia, lymphatics and lymph nodes like ileocolic nodes, pericolic nodes, nodes at the original of SMA.
   - If the growth is fixed to posterior abdominal wall, common iliac vessels, palliative ileotransverse anastomosis is done (ileotransverse colostomy).
2. **Carcinoma transverse colon**—'V' resection. The area supplied by middle colic artery is removed followed by end to end anastomosis. The patient may need removal of entire transverse colon depending upon lesion.
   - When lesion is at hepatic flexure or in the transverse colon, extended right hemicolectomy should be done.
3. **Carcinoma left colon**—left radical hemicolectomy.
   - Left half of the transverse colon and descending colon are removed followed by anastomosis of transverse colon to sigmoid colon—This is the area supplied by left colic artery (Fig. 29.37).

KEY BOX 29.12

STRUCTURES THAT CAN GET INJURED DURING RIGHT HEMICOLECTOMY

- Duodenum
- Ureter
- Gonadal vessels

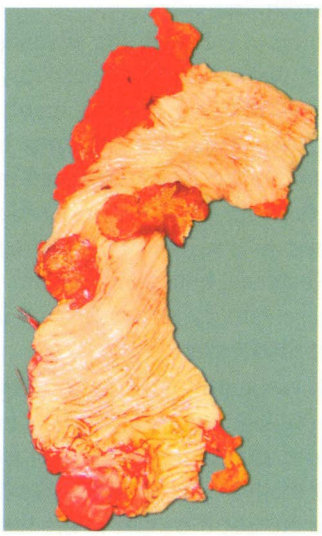


Fig. 29.36: Right radical hemicolectomy

[1]Removal of a foot of ileum is not necessary unless it has a doubtful vascularity after ligation of pedicles.

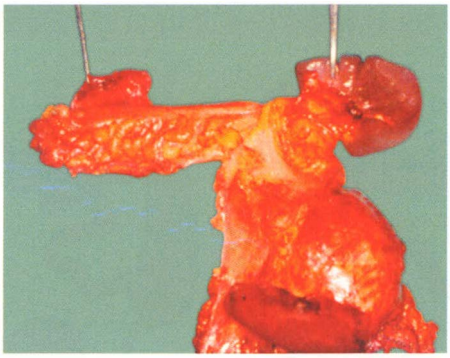


Fig. 29.37: Left radical hemicolectomy including removal of the spleen as the growth was infiltrating the spleen—routine removal of spleen should not be done

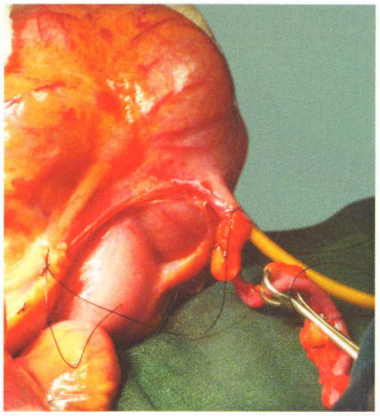


Fig. 29.38: 'On-table' irrigation through appendicular stump

4. **Carcinoma sigmoid colon**—radical sigmoid colectomy followed by anastomosis of descending colon to the rectum (colorectal anastomosis). Or in a few cases, left hemicolectomy may have to be done.
5. **Left-sided colonic tumours with intestinal obstruction**—an emergency temporary transverse colostomy is done to divert the faecal matter and to relieve intestinal obstruction (for more details on colostomy *see* page 785). Resection is not done because many patients are elderly with comorbid illness such as diabetes, hypertension, cardiac illness.
   - General condition of the patient is poor with gross abdominal distension and dehydration.
   - Left colon is loaded with faecal matter. Hence, high chances of anastomotic leakage and faecal peritonitis are present.
   - After 2 weeks, laparotomy is done once again. The primary tumour is resected and end-to-end anastomosis done.
   - This is followed by closure of the colostomy 8 weeks later—**three-stage operation.**
   - Single-stage resection can also be done provided, thorough colonic irrigation through appendicular stump (after appendicectomy), is given and it should be successful in cleaning the entire colon. This is the concept of on-table irrigation and lavage.

ON-TABLE IRRIGATION AND LAVAGE (Fig. 29.38)

Indication

In cases of left-sided colonic obstructions—classical example being carcinoma rectosigmoid with obstruction.

Procedure

- Resection is done first. Clamps are applied to both ends.
- Appendicectomy is done and purse string suture applied but not tied.
- Through the appendicular stump lumen, a 30 Fr Foley catheter is passed into colon.
- The position of the catheter is checked to be lying safely in the caecum and its balloon is inflated. A purse string suture, previously applied to the appendix base is tied.
- Saline is irrigated at the rate of 50 to 100 ml per hour speed. Proximal clamp is opened into a container (kidney tray).
- It takes about an hour or so for the whole gut irrigation.
- This is done till the returning fluid is clear.

Advantage

It avoids a stoma, decreases stay in the hospital and thus less expensive.

Caecostomy tube

Once anastomosis is completed, Foley catheter is brought out through an opening in the abdominal wall and connected to a bag. It is sutured to the inside of the parietes by vicryl sutures and kept open.

Tube removal

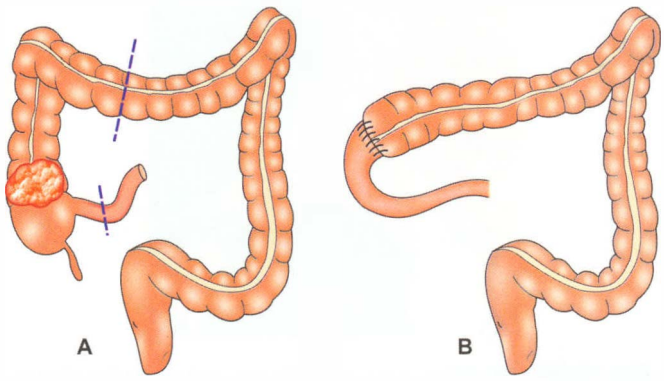
Tube is removed after 7–10 days provided there is no leak from the suture line.

ENHANCED RECOVERY PROGRAMME

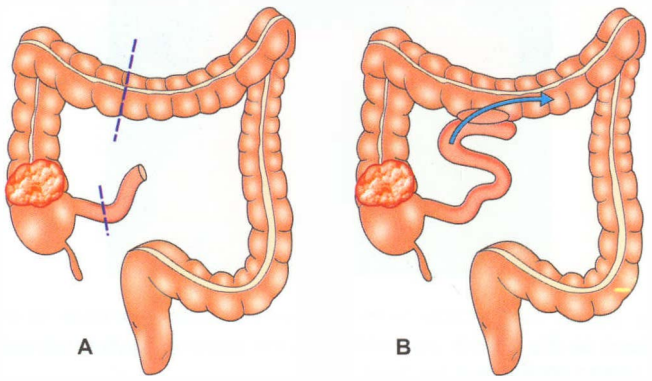
These are the various changes in the management of these patients over conventional methods. A few are given below which will help the patient recover fast and get discharged—no quality compromise.

- Nasogastric tube is not required unless operating on obstructed colons. Evidence is that the sutures /staplers are good enough to prevent the leaks.
- Improve the nutrition preoperatively by increased intake of carbohydrates—oral or TPN.
- Laparoscopy has the definite advantage of small incisions, less pain, easy breathing and less respiratory complications.
- Early mobilization, early feeds, early recovery and early removal of catheters.
- Epidural catheters to relieve pain.
- Few diagrammatic representation of the colectomies are shown in the next page (Figs 29.39 to 29.44).

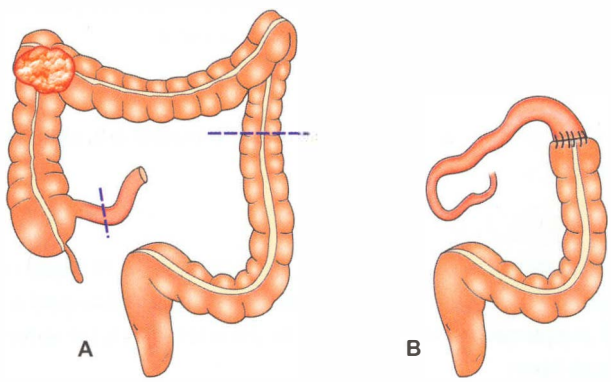
SURGERIES IN A CASE OF CARCINOMA COLON (Figs 29.39 to 29.44 and Table 29.4)



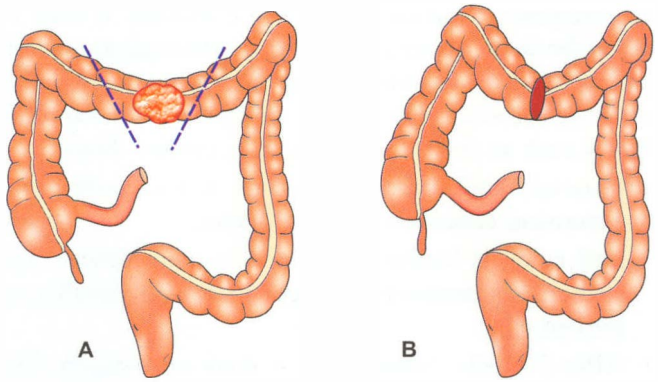
Figs 29.39A and B: Right hemicolectomy followed by end-to-end anastomosis—ileocolic



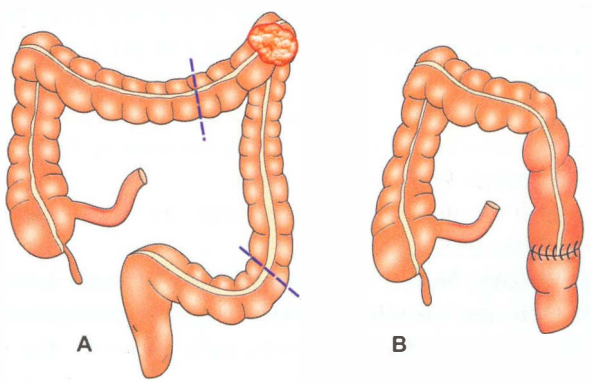
Figs 29.40A and B: Palliative ileotransverse anastomosis inoperable case



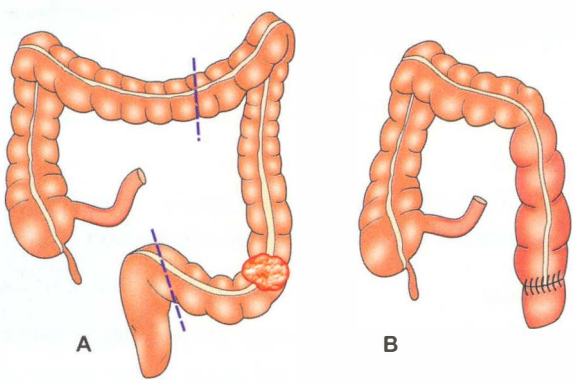
Figs 29.41A and B: Extended right hemicolectomy—growth in the hepatic flexure



Figs 29.42A and B: Transverse colectomy followed by colocolic anastomosis



Figs 29.43A and B: Left hemicolectomy for growth in the splenic flexure



Figs 29.44A and B: Resection of rectosigmoid growth followed by colorectal anastomosis

Figs 29.39 to 29.44: All figures are contributed by Ms Vidushi Kapil, MBBS student, KMC, Manipal

Table 29.4 Summary of the resections for carcinoma colon

Site of the tumour	Name of surgery	Part of the bowel resected	Artery supplying	Safe margin
Caecum	Right hemicolectomy	Terminal ileum to midtransverse colon	Ileocolic, right colic RIGHT branch of middle colic	5 cm
Ascending colon	Right hemicolectomy	Terminal ileum to midtransverse colon	Ileocolic, right colic RIGHT branch of middle colic	5 cm
Hepatic flexure	Extended right hemicolectomy	Terminal ileum to descending colon	Ileocolic, right colic MIDDLE colic	5 cm
Transverse colon	Extended right hemicolectomy	Terminal ileum to descending colon	Ileocolic RIGHT colic MIDDLE colic	5 cm
Splenic flexure	Extended left hemicolectomy	Right flexure to rectosigmoid	Middle colic LEFT colic IMA	5 cm
Descending colon	Left hemicolectomy	Left flexure to sigmoid	IMA LEFT branch of middle colic	5 cm

POSTOPERATIVE CHEMOTHERAPY

1. pT1-2N0M0 do not require any adjuvant treatment, such patients can be kept on follow-up with routine 3 monthly CEA and annual CECT thorax/abdomen/pelvis.
2. pT3N0M0 or node positive disease requires adjuvant treatment in the form of concurrent chemoradiotherapy and chemotherapy. 2 cylces of FOLFOX (5-FU + Leucovorin + Oxaliplatin) → Concurrent 5-FU/Leucovorin and radiation → 2 more cycles of FOLFOX.
   Oral Capecitabine can be used in place of IV 5-FU.
3. It is preferable to add Oxaliplatin in the chemotherapy regimen if nodes are positive for metastatic disease. In older population (> 65–70 years), it might of less benefit.
4. Oxaloplatins have been shown to down size liver metastasis. Chief complications of Oxaliplatins is peripheral neuropathy.

- Indications for chemotherapy has been given in Key Box 29.13.

KEY BOX 29.13

INDICATIONS FOR CHEMOTHERAPY

- All node positive patients
- In node negative patients if
  - T4 lesions are involving free mesothelial surface
  - Major microscopic vein involvement
  - **Signet cell carcinoma**
  - High preoperative CEA
  - Aneuploidy on flow cytometry
  - Microsatellite instability

Postoperative radiotherapy

Adenocarcinoma colon does not respond well to radiation. Routinely it is not given. Surgery remains the gold standard for carcinoma colon. Soft tissue infiltration into psoas muscle or abdominal wall or inoperable recurrent tumours are indications for radiotherapy.

Metastatic disease without obstruction

Patients with isolated liver/lung secondaries should also undergo treatment with a radical approach as even in these cases with resection of the primary and adequate liver/lung resection, a good disease control can be achieved.

1. A typical course of neo-adjuvant therapy comprises concurrent 5-FU/Capecitabine and radiation in cases of large lesions abutting the abdominal wall or down into the pelvis. A dose of 45–50 Gy is used to treat the pelvis including the growth and the draining lymphatic regions followed by 5 Gy boost to the tumour itself.
2. Following neo-adjuvant therapy, patient should be re-evaluated using CT/MRI for possibility of resection.
3. Surgery is usually considered after 6–8 weeks following neo-adjuvant therapy as the maximal response to the treatment may take up to 2 months.
4. Further adjuvant treatment is to be given following surgery depending upon the histopathological report.

Management of liver secondary

- CT scan and PET scan are done to evaluate local/systemic disease. Provided there is no systemic spread, liver secondaries have to be treated aggressively. Pattern of recurrence in colonic carcinomas is more commonly distant, that is they tend to recur more commonly at distant sites such as liver, and lungs. As a result, systemic treatment is more necessary.
- Liver directed therapies such as hepatic arterial chemotherapy infusion/embolisation, radiofrequency-ablation, radiotherapy should be used in treatment of isolated liver metastasis.
- Indications for liver resection is given in Key Box 29.14.

KEY BOX 29.14

INDICATION FOR RESECTION OF LIVER METASTASIS

- Solitary metastasis or metastasis confined to one lobe.
- < 3 metastasis in both lobes
- Absence of extrahepatic disease

Novel agents in colorectal cancers

1. Bevacizumab—anti-VEGF (vascular endothelial growth factor) monoclonal antibody. It has anti-angiogenesis property thereby controlling the tumour growth.
2. Cetuximab anti-EGFR (epidermal growth factor) monoclonal antibody.

Follow-up (Key Box 29.15)

Most of the colonic cases are curable if diagnosed and treated early. Also metachronous lesion can occur in the rest of the colon. Hence, certain tests are necessary during follow-up.

Treatment of recurrent or metastatic cancer

- Recurrence or metastasis is suspected during follow-up by abnormal values of investigation.
- Recurrent tumour should be resected *en bloc*—it may amount to a more radical procedure including resection of duodenum, liver, kidney (Figs 29.45 and 29.46)
- Metastasis in the liver (Key Box 29.14).

KEY BOX 29.15

FOLLOW-UP OF COLORECTAL CANCER

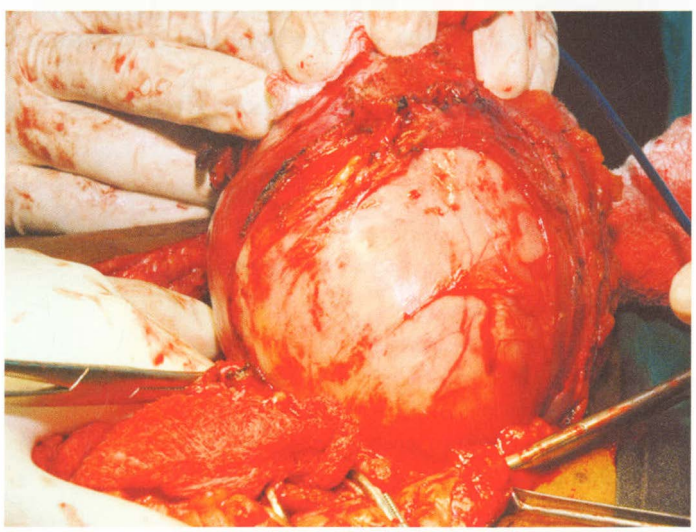
Tests	Duration	In years
1. Haemo-occult	Once in 3–6 months	3
2. Colonoscopy	6 months after surgery, later once in a year	3
3. Alkaline phosphatase	Once in 3–6 months for 3 years	3
4. CEA	Once in 6 months	3
5. Chest X-ray	Yearly	

CHEMOPREVENTION OF COLONIC CANCER

1. **Folic acid:** It is an important vitamin with many functions. In the absence of folic acid, hypomethylation can occur. As a result of this overexpression of proto-oncogenes such as K-ras and c-Myc can occur. Deficiency of folic acid causes imbalances in the nucleotide pool leading to DNA break and mutation. Thus folic acid supplementation should be given in adenoma specially when baseline levels of folic acid is low.
2. **Dietary fibres:** Fibres decrease the transit time , they dilut the carcinogens and are used to present development o cancer. Cellulose, hemicelluloses and pectin are a few examples. Fibres also produce short chain in fatty acid causing fermentation by faecal flora. Thus colonic pH becomes more acidic which in turn inhibits carcinogenesis
3. **Aspirin, calcium, and sulindac** also have been used to prevent cancer developing in an adenoma.

COLON SCREENING

- Large bowel is the 4th most common site for cancer after lung, stomach and breast
- More common in North America, North Europe and Australia. Lowest rates in Africa, India.
- 75% of CRC develop in people with no known risk factors apart from older age.



Figs 29.45 and 29.46: A colorectal cancer patient who was operated 3 years back for carcinoma caecum presented with recurrence in the abdominal wall. He underwent excision of the tumour. He did not have any other site of metastasis. Postoperative radiotherapy was given

Screening options

1. Faecal occult blood test (F●BT)

- It is guaiac test which will detect elevated level of blood in stool. It requires 2 samples from each of 3 consecutive stools which are smeared onto cards.
- **False positive:** Vegetables, fruits, red meat, aspirin or any other bleeding lesion proximal to colon screening, FOBT has shown to decrease mortality by 20 to 30%.

2. Flexible sigmoidoscopy (FS)

- Reduces incidence and mortality of distal CRC by around 60%.
- Single FS at the age of 60 is recommended in UK.
- In US, 5-yearly screening is being done.

3. Colonoscope screening

- Should be done if there is distal adenoma (chances of proximal adenoma are high).
- 70% of all advanced colorectal neoplasia will be detected with this strategy.
- Procedure is painful, requires sedation and analgesics.
- Chances of perforation are 1 in 500 to 2000 cases.
- It requires skills of an experienced endoscopist.

4. Virtual colonoscopy

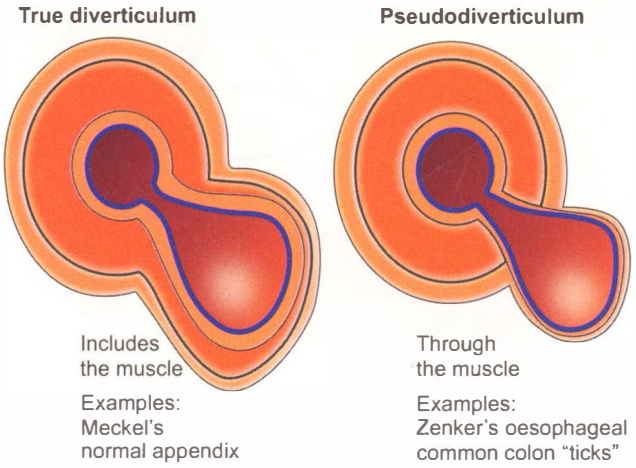
- It is an alternative but not yet become popular because of time, cost and preparation.
- It is done with the help of CT scan.
- Biopsy cannot be taken

DIVERTICULAR DISEASE OF COLON

It is an acquired condition, in which colonic mucosa herniates through the circular muscle fibres at weak points, where blood vessels penetrate the colonic wall. Since it is acquired it lacks the muscle coat. They are thin, more prone for infections and perforation. Hence, they are termed pseudodiverticuli.

Aetiopathogenesis

- The disease is common in western population wherein diet is very **poor in fibres** because of refining of sugar and flour. **Nonstarch polysaccharides (NSP)** or low dietary fibres are the chief factor for **diverticulosis of colon** (Key Box 29.16).
- It requires very high pressure for propulsion of faecal matter and this is believed to cause characteristic thickening of muscles and herniation of mucosa (Figs 29.47 and 29.48).
- In Africans and Indians, the disease is rare because of high fibre content of the diet.
- The disease starts after the age of 40. Any stress or emotional disorders may add to the constipation already caused by dietary factors and result in diverticular formation.
- **90% of them affect sigmoid colon.** Rectum is spared in majority. Rarely, it affects right colon.



Figs 29.47 and 29.48: Diverticulum

- Diverticulae project between antimesenteric and mesenteric borders with taenia but they never penetrate taenia (Fig. 29.49).
- There is **muscle hypertrophy,** which project into the lumen as obstructive folds. The mucosa is essentially normal. Slowly, luminal diameter is narrowed.
- **Inflammation** occurs in the pericolonic tissue with or without abscess formation.

KEY BOX 29.16

NSP AND DISEASES

- Diverticular disease
- Obesity and diabetes mellitus
- Constipation, and piles
- Breast cancer and colonic cancer

Structural changes in colonic wall of patients with diverticulosis

- Mycosis
- Thickening (neither hypertrophy nor hyperplasia) of the circular muscle layer.
- Shortening of the taenia coli
- Luminal narrowing
- ↑ elastin deposition in taenia coli
- ↑ type III collagen synthesis
- ↑ collagen cross-linking

Segmentation

- Law of Laplace: Pressure = K × Tension/Radius
- Sigmoid colon has small diameter resulting in highest pressure zone.
- Segmentation = motility process in which the segmental muscular contractions separate the lumen into chambers.
- Segmentation →increased intraluminal pressure → mucosal herniation →diverticulosis.

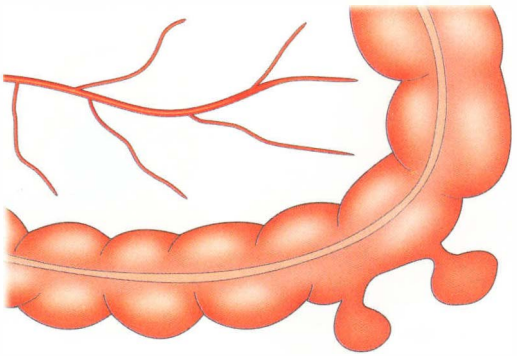


Fig. 29.49: Sigmoid diverticulae

- May explain why high fibre prevents diverticuli by creating a larger diameter colon and less vigorous segmentation.
- Collagen—connective-tissue diseases such as Ehlers-Danlos syndrome, Marfan's syndrome, and autosomal-dominant polycystic kidney disease result in structural changes in the bowel wall, leading to decreased resistance of the wall to intraluminal pressures and thus allowing protrusion of diverticula.

Clinical features

1. **Diverticulosis:** It refers to presence of diverticulosis without much symptoms. But on careful questioning, patients do have lower abdominal distention, heaviness, flatulence, etc. Vague abdominal pain is also felt in the left iliac fossa.
2. **Diverticulitis:** Left-sided lower abdominal pain, moderate to severe is associated with passage of loose stools. The pain is partially relieved on passing flatus.
   - Bleeding per rectum can be the presenting feature, sometimes it can be massive.
   - Low-grade fever, tenderness, rigidity and even mass may be present in the left iliac fossa (like left-sided appendicitis). The mass is thickened, inflamed, tender and sigmoid. Such attacks result in abscess which rupture into hollow organs and give rise to fistulae (Fig. 29.50).

Fig. 29.50: Perforated diverticula at surgery—resected specimen (*Courtesy:* Dr Ramesh Rajan, Surgical Gastroenterologist, Trivandrum Medical College, Kerala)

3. **Internal fistulae:** Colovesical fistulae give rise to pneumaturia (flatus in the urine) and rarely faeces in the urine (Key Box 29.17).

KEY BOX 29.17

CAUSES OF INTERNAL FISTULAE

- Diverticular disease of colon
- Carcinoma of colon
- Crohn's disease
- Radiation
- Tuberculosis

For summary of diverticular disease *see* Fig. 29.51

Classification/staging system

Hinchey classification (Fig. 29.52)

I. Pericolic abscess
II. Walled off 3 pelvic abscess
III. Generalised purulent peritonitis
IV. Generalised faecal peritonitis

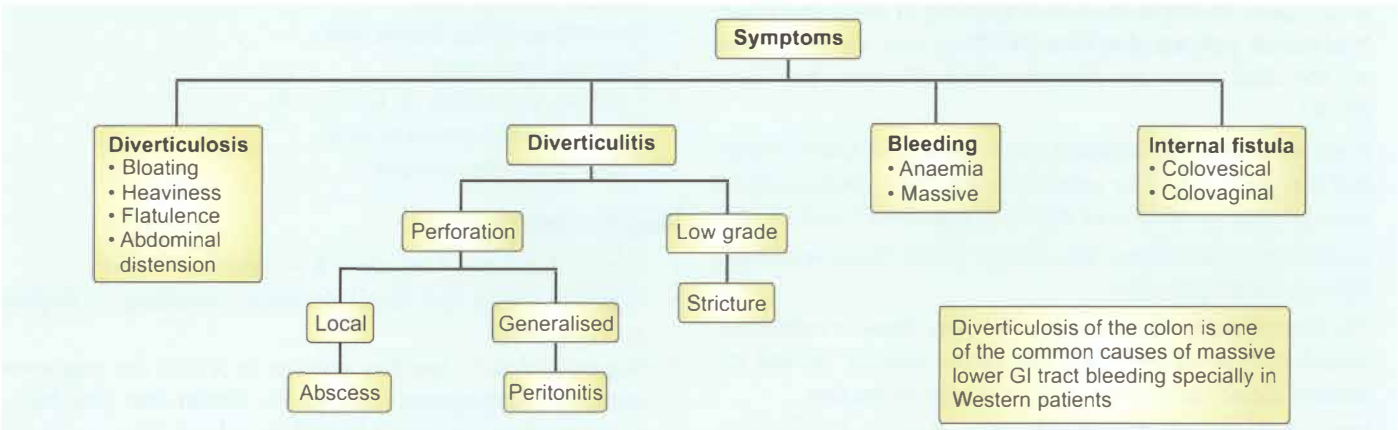


Fig. 29.51: Summary of symptoms of diverticular disease

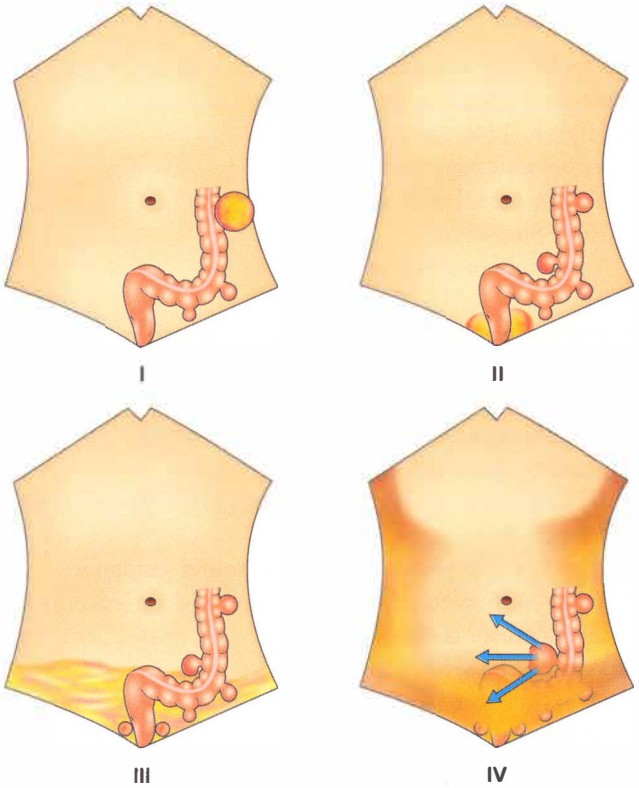


Fig. 29.52: Hinchey classification

Hinchey stages I and II may be treated by sigmoid colectomy and primary anastomosis (a one-stage operation).

Hinchey stages III and IV are treated by sigmoid colectomy followed by end-colostomy and Hartmann pouch.

PEARLS OF WISDOM

The most common fistula in acute diverticulitis is colovesical followed by colovaginal fistula.

Investigations

1. **Sigmoidoscopy:** Mucosa may be normal or may show erythematous and oedematous changes. Ulcers are absent. Opening of diverticulae can be seen.
2. **Barium enema: Contraindicated in acute cases.**
   - It may show **saw-tooth** appearance due to muscle hypertrophy.
   - It may show a long stricture
   - **Champagne glass sign:** Partial filling of diverticula by barium with stercolith inside the diverticula.
3. **Colonoscopy** to confirm the findings and to rule out **carcinoma colon** (Figs 29.53 and 29.54).

PEARLS OF WISDOM

Sigmoidoscopy, colonoscopy and barium enema are contraindicated in acute diverticulitis.

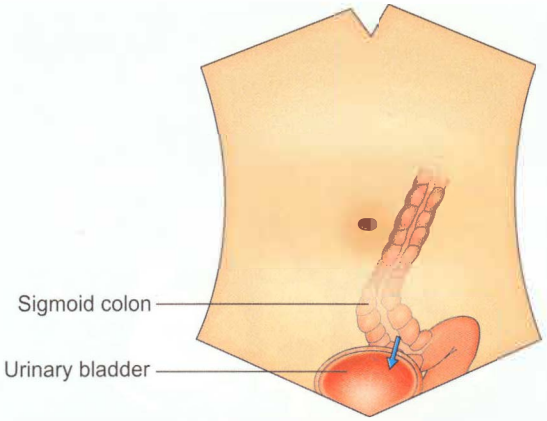


Fig. 29.53: Colovesical fistula (arrow)

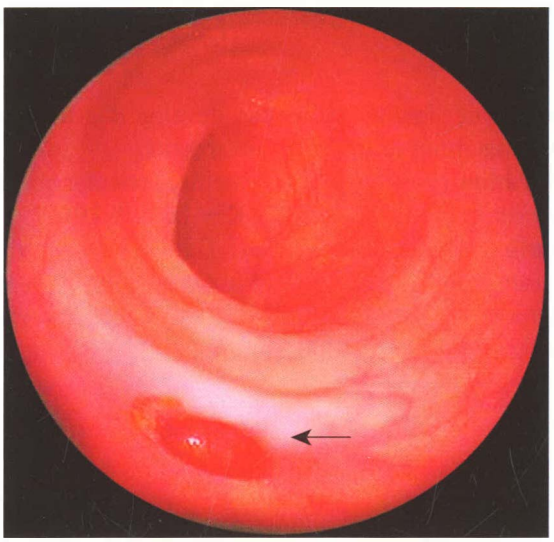


Fig. 29.54: Colonoscopy showing opening of diverticula

4. Ultrasound and CT scan are the investigations of choice in acute diverticulitis (Fig. 29.55).

COMPLICATIONS

1. **Massive haemorrhage** per rectum: Haemorrhage is due to vessels in the base of diverticulae, more so in atherosclerotic or hypertensive patients.

KEY BOX 29.18

CT SCAN ACUTE DIVERTICULITIS

- Can detect thick muscular folds—confirms the diagnosis.
- Detects an abscess and can confirm complication.
- Detects extraluminal air or contrast—confirms perforation.
- Can rule out other causes—acute pancreatitis with pericolic collection, etc.
- **It is the investigation of choice in acute diverticulitis.**
- Thickened colonic wall > 4 mm
- Pelvic abscess can be diagnosed

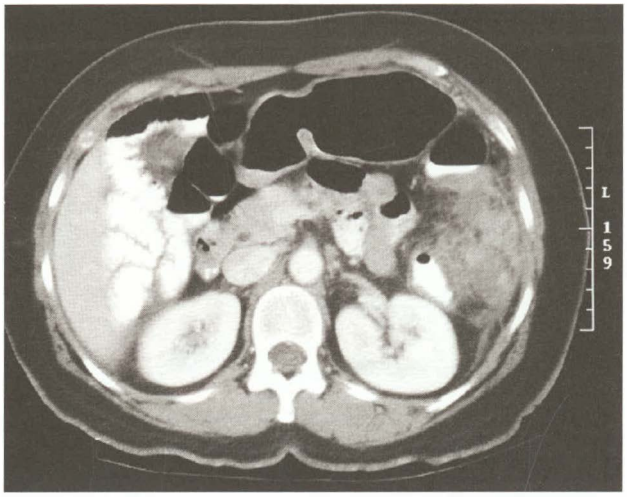


Fig. 29.55: CT scan showing pericolic abscess on the left paracolic space

2. **Stricture** of sigmoid colon can develop due to recurrent attacks resulting in intestinal obstruction.
3. **Perforation** may result in peritonitis, pericolic abscess or pelvic abscess (Fig. 29.56 and Key Box 29.19).
4. **Fistula formation:** Internal fistulae occur due to inflammatory adhesions and abscess formation which ruptures resulting in fistula. Thus, colovesical, colovaginal, colointestinal fistula can occur.

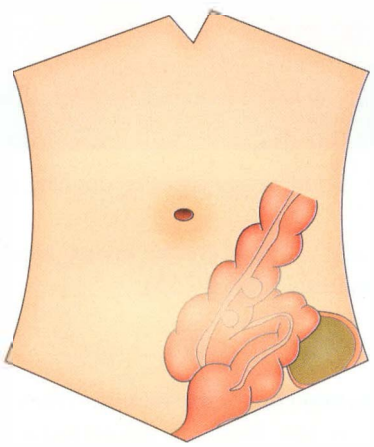


Fig. 29.56: Pericolic abscess

Differential diagnosis

- Carcinoma of the colon
- Inflammatory bowel disease
- Ischaemic colitis
- Irritable bowel syndrome
- Pelvic inflammatory disease

Indications for surgery

1. Failure to respond to conservative/dietary advice
2. 2 attacks of diverticulitis
3. Complications

TREATMENT

1. **Stage of diverticulosis** or in those patients who have recovered from one attack of diverticulitis.
   - High residue diet
   - Fruits and vegetables
   - Whole meal bread and flour
   - Bulk purgative
   - To avoid constipation

   Diet
   - High fibre diet, optimal amount of daily fibre is unknown.
   - 20 to 30 g per day is a widely recommended.
   - Recommendation to avoid seeds, nuts and popcorn.
2. **Acute diverticulitis with pericolic abscess**
   - Rest, hospitalisation, correct hydration
   - IV antibiotics: Bactericidal against gram –ve and anaerobes.
   - Abscess is aspirated under ultrasound guidance.
   - After 4–6 weeks, elective sigmoid colectomy and anastomosis is done.
3. **Diverticulitis with peritonitis**

   A. **Hartmann's procedure** is the choice: Sigmoid colon is resected, end-colostomy is done by using descending colon followed by closure of rectal stump (Figs 29.57A and B).
   - After 4–6 weeks, colorectal anastomosis is done.

   B. However, if a **perforation is small** and the general condition is good after the resection, the colon is irrigated with 8–10 litres of saline till the contents are clear. This is followed by colorectal anastomosis in the same sitting.

KEY BOX 29.19

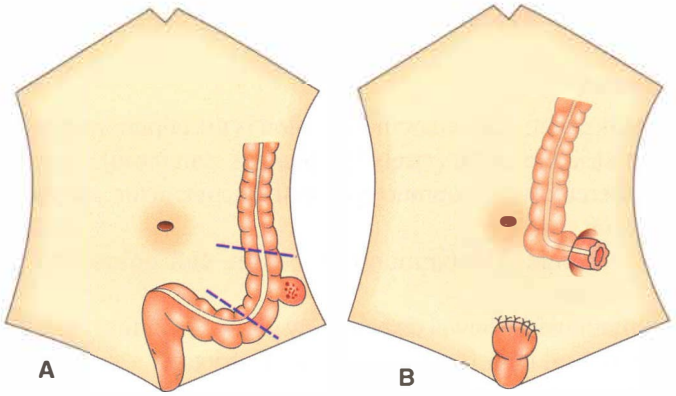
PERICOLIC ABSCESS

- Abscess depends on the ability of the pericolic tissues to localise the spread of the inflammatory process.
- Intra-abdominal abscesses are formed by—anastomotic leakage: 35%—diverticular disease: 23%.
- Limited spread of the perforation forms an inflammatory phlegmon, while further (but still localised) progression creates an abscess.
- Signs and symptoms: High grade fever with or without leukocytosis despite adequate antibiotics, tender mass.
- Treatment:

A. Small pericolic abscess: 90% will respond to antibiotics and conservative management alone. Often it is retrocolic. May present as swelling in the loin.

B. Percutaneous abscess drainage (PAD) is the treatment of choice for small, simple, well-defined collections. 100% success in simple unilocular abscesses.

C. Open drainage: In cases of multilocular collection, abscesses associated with enteric fistulas, and abscesses containing solid material. Drainage of pus, resection, Hartmann's operation is ideal choice. Closure of the colostomy after 6–8 weeks is done.



Figs 29.57A and B: Hartmann's procedure

4. **Treatment of fistulae:** As an elective procedure, with good preparation, after confirming the site of fistula, resection of the sigmoid colon with closure of fistula can be done.

KEY BOX 29.20

TYPES OF FISTULA RELATED TO DIVERTICULAR DISEASE

- Colovesical: 65%
- Colovaginal: 25%
- Colocutaneous
- Coloentero

MISCELLANEOUS

FAECAL FISTULA

Classification (Key Box 29.20)

Depending upon the nature of the disease or a surgical procedure.

I. **Primary or type I fistula:** It develops as a result of an underlying disease affecting gut. Example: Carcinoma colon infiltrating urinary bladder resulting in colovesical fistula or rupture through the skin resulting in pericolic abscess, fistula.

II. **Secondary or type II fistula:** Occurs after injury to otherwise normal gut. Examples: Left colonic/right colonic injury in percutaneous nephrolithotomy (PCNL) for renal stones or following anastomotic leak.

Depending upon the site of the fistula

A. **Lateral fistula:** It occurs after a colocolic or ileocolic anastomosis wherein a few sutures would have given way in the immediate postoperative period.

B. **End fistula:** This means both ends of the intestines are open following resection and anastomosis giving rise to postoperative peritonitis.

Bacterial flora

- More than 99% of faecal bacterial flora are **anaerobic**.
- Most common anaerobe is ***Bacteroides fragilis,*** with a count of 10^{10}/g of wet faeces. Clostridia, Lactobacillus are other organisms.
- ***E. coli*** is the predominant **aerobic organism**. Count is 10^{7}/g of faeces. Klebsiella, Proteus, Enterobacter are other **aerobes**. ***Streptococcus faecalis*** is the principal enterococcus.

Clinical manifestations

- Postoperative patient 'not doing well'—prolonged paralytic ileus, faeculent discharge from wound or drainage site, etc. is an indication of faecal fistula.
- Features of septic shock: Often patient may not complain of abdominal pain but manifestations can be of renal failure, tachycardia, tachypnoea and hypotension.

Diagnosis

- Total counts are elevated—it indicates infection.
- Creatinine and urea values are high indicating renal failure.
- Ultrasound to detect any collection.
- CECT can detect intraperitoneal collection, pneumoperitoneum, anastomotic leak. However, creatinine should be normal before doing CT scan. Often patients are in sepsis with renal failure.

Treatment (Figs 29.58 to 29.60)

- Control of infection
- Drainage of sepsis—laparotomy, resection and anastomosis, ileostomy or colostomy may be necessary.
- Lateral fistula may heal with total parenteral nutrition provided there is no distal obstruction (TPN).
- End fistula may require resuturing with or without proximal diversion colostomy/ileostomy.`

Fig. 29.58: This patient underwent three surgical procedures for treatment of fistula. Initial surgery was right hemicolectomy for ileocaecal tuberculosis. Second surgery was resection and a proximal diversion ileostomy because of the leak and finally after two months, closure of ileostomy was done followed by ileotransverse anastomosis

Fig. 29.59: Ileostomy following leak in the case mentioned in Fig. 29.58

Fig. 29.60: A case of faecal fistula following ileal resection for obstruction caused by a band. Following resection anastomosis on the fifth postoperative day air mixed faecal matter started coming out from the wound. CT scan revealed small lateral fistula. The patient responded to conservative treatment with TPN given for fourteen days. Fistula subsided

COLONIC STRICTURE

Causes

1. Malignant: Adenocarcinoma colon is the commonest cause of stricture colon (rarely lymphomas, carcinoid).
2. Tuberculosis: Uncommon cause of stricture in the ascending colon.
3. Ischaemic: Uncommon/rare cause—left colon may be affected.
4. Inflammatory bowel disease: Any ulcers, including amoebic, may heal with fibrosis resulting in stricture.
5. Diverticular stricture
6. Radiation stricture
7. Endometriomas: Ectopic endometriosis tissue responds to cyclic hormonal stimulation causing inflammation and fibrosis.

Clinical features

- Progressive constipation
- Change in bowel habits
- Bleeding per rectum
- Features of large bowel obstruction
- Mass may/may not be felt

Investigation

Colonoscopy and biopsy

Treament

- Single-stage resection and end-to-end anastomosis.
- Treatment of the cause

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated.
- More and more cases of large bowel obstruction are treated by single-stage resection after giving an 'on-table' lavage.
- Role of chemotherapy for carcinoma has been updated.
- Faecal fistula, colonic stricture have been added.
- Chemoprevention of colonic cancer is added.
- Enhanced recovery programme has been added.
- Ten commandments of colonic resection have been added.

MULTIPLE CHOICE QUESTIONS

1. The following feature is true of adenomatous polyps of the large intestine:
A. Most are sessile
B. Most can be removed by colonoscopic snaring
C. Adenoma smaller than 15 mm in diameter do not carry the risk of malignant potential
D. Young people are more likely to have these polyps

2. Common premalignant conditions for colonic cancer include the following *except*:
A. Familial polyposis coli
B. Ulcerative colitis
C. Adenomatous polyp
D. Peutz-Jeghers syndrome

3. Flexion of the hip can be present in:
A. Carcinoma splenic flexure
B. Carcinoma caecum
C. Carcinoma sigmoid colon
D. Carcinoma hepatic flexure

4. Abscess in the lateral abdominal wall can be a feature of:
A. Acute pancreatitis
B. Perforated carcinoma caecum
C. Diverticular perforation
D. Meckel's diverticular perforation

5. Mechanical bowel preparation is best given using:
A. Plenty of saline
B. Oral mannitol
C. Glycerine enema
D. Whole gut irrigation using oral polyethylene glycol

6. Which one of this is true in colonic surgery?
A. No touch technique of Turnbull
B. Right-sided lesions are treated by colostomy
C. Left-sided lesions are treated by resection
D. Removal of 30 cm of ileum along with colon

7. Following is true about prognostic factors of carcinoma colon *except:*
A. Elderly patients have poorer prognosis
B. Perioperative blood transfusion has poor prognosis
C. Survival is good if it is limited to mucosa and there are no nodes
D. Obstruction and perforation is associated with poor prognosis

8. Regarding colonoscopic screening, which one of the following is true?
A. It is done if there is distal adenoma
B. It is painless and easy to perform
C. It is not useful for screening of colonic cancer
D. Detection of all advanced colorectal neoplasia is only up to 20%

9. Proliferative growth is more common in the _____ colon:
A. Right
B. Left
C. Rectosigmoid
D. Splenic flexure

10. The 'Gold standard' investigation for detection of colonic cancer is:
A. Ultrasound abdomen
B. CT scan
C. Barium enema
D. Colonoscopy and biopsy

11. Following are the features of intestinal tuberculosis *except:*
A. It can be secondary to pulmonary tuberculosis
B. Terminal ileum and caecum are commonly involved
C. Ulcers are transverse
D. Ulcers do not result in structure

12. In which of the following malignancy anaemia is an important method of presentation?
A. Malignant melanoma
B. Carcinoma breast
C. Carcinoma caecum
D. Carcinoma pancreas

13. The most common fistula in diverticulitis is:
A. Colovesical
B. Colovaginal
C. Colorectal
D. Colocolic

14. Which of the following diverticula is true diverticula?
A. Sigmoid diverticuli
B. Meckel's diverticuli
C. Parabronchial diverticulum
D. Laryngeal diverticula

15. In hereditary nonpolyposis colorectal cancer, which carcinoma is more often seen?
A. Rectum
B. Rectosigmoid
C. Transverse colon
D. Caecum and ascending colon

16. Incidence of malignancy in familial polyposis coli is:
A. 10%
B. 30%
C. 50%
D. 100%

17. Following are true for carcinoembryonic antigen *except*:

A. It is a glycoprotein
B. It is tumour marker for carcinoma colon
C. It should be done in all cases of carcinoma colon before surgery
D. Produced by colorectal epithelium and cleared by kidney

18. Following are true for right hemicolectomy for carcinoma caecum *except:*

A. Right one-third of transverse colon is also removed
B. Greater omentum should be removed
C. Terminal 30 cm of the ileum should be removed
D. Duodenum can get injured during surgery

19. Indications for postoperative chemotherapy following colectomy for carcinoma include following *except*:

A. Signet ring carcinoma
B. Lymph nodes are positive
C. Lymphovenous involvement
D. Involvement of muscularis propria

20. The most common aerobic organism present in a sigmoid colonic faecal fistula is:

A. Clostridia
B. Lactobacillus
C. *Bacteroides fragilis*
D. *E.coli*

ANSWERS

1 B	2 D	3 B	4 D	5 D	6 A	7 A	8 A	9 A	10 D
11 D	12 D	13 A	14 B	15 D	16 D	17 D	18 C	19 D	20 D

30 Intestinal Obstruction

- Sigmoid volvulus
- Meckel's diverticulum
- Adhesions and bands
- Gall stone ileus
- Intussusception
- Mesenteric vascular occlusion
- Hirschsprung's disease
- Atresia and stenosis
- Arrested rotation with bands
- Volvulus neonatorum
- Meconium ileus
- Imperforate anus
- Food bolus obstruction
- Paralytic ileus
- Malrotation and midgut volvulus
- What is new?/Recent advances

Introduction

- Intestinal obstruction is a challenging surgical emergency encountered by general surgeons. This can affect any age group starting from neonate to an old man. It can affect a school going boy, working woman or a man during their peak of life. Sometimes it can be fatal either due to delay in the diagnosis, delay in the treatment or complications related to surgery. Abdomen is a Pandora's box. Sometimes, it is difficult to pinpoint the cause of obstruction.
- Adhesions and hernia are the two most common causes of intestinal obstruction. Adhesions are more common than hernias nowadays. Laparoscopic surgery has definitely decreased incidence of adhesions. In Western countries more than 50% cases of intestinal obstruction are due to adhesions and only 10–15% are due to obstructed hernia (Fig. 30.1). However, students should be able to diagnose intestinal obstruction, resuscitate the patients and refer the patient for further surgical treatment. With the availability of sophisticated investigations such as CT scan, diagnosis can be established in majority of cases before surgery. However, in other cases, **'exploratory laparotomy'** will give the diagnosis.

Definition

When the intestinal contents fail to move distally, it is called intestinal obstruction. It is the most common surgical disorder (emergency) of the intestines.

Few important facts about intestinal obstruction:

- 80% occur in small bowel
- 20% occur in large bowel
- Majority (more than 80%) of small bowel obstructions are benign in nature.

In the large bowel, more than 70% of colonic obstruction is due to malignancy—others being inflammatory bowel diseases, ileocaecal tuberculosis, volvulus, etc.

CLASSIFICATION

I. Depending upon the nature of obstruction

(Key Box 30.1)

A. Dynamic obstruction/mechanical obstruction (Fig. 30.1)
B. Adynamic obstruction—paralytic ileus or neurogenic ileus.

Fig. 30.1: Obstructed incisional hernia (*Courtesy:* Dr CG Narasimhan, Senior Consultant Surgeon, Mysore, Karnataka)

KEY BOX 30.1

COMMONLY USED TERMINOLOGY

- **Mechanical obstruction**
  There is a physical barrier which prevents the abnormal progress of intestinal contents.
- **Paralytic ileus**
  There is no physical barrier but failure of peristalsis to propel intestinal contents due to neurogenic causes.
- **Simple obstruction**
  It refers to obstruction to lumen only (early cases)
- **Strangulated obstruction**
  It refers to obstruction with impairment of blood supply to the gut.
- **Closed loop obstruction**
  In this condition, the intestine is occluded in two places. More chances of gangrene and perforation are present, e.g. volvulus.
- **Pseudo-obstruction:** No mechanical cause.

II. Depending on the blood supply

A. Simple obstruction: Blood supply is not seriously impaired.

B. Strangulated obstruction: Blood supply is seriously impaired.

C. Closed loop obstruction: It means both proximal and distal ends are blocked. This occurs in carcinoma of the right colon with constrictive lesions. If the ileocaecal valve is competent and the obstruction is total, the intraluminal pressure within the colon increases. As a result of this, the caecum may perforate. Thus, closed loop obstruction can be dangerous (Fig. 30.2). Another example is sigmoid volvulus.

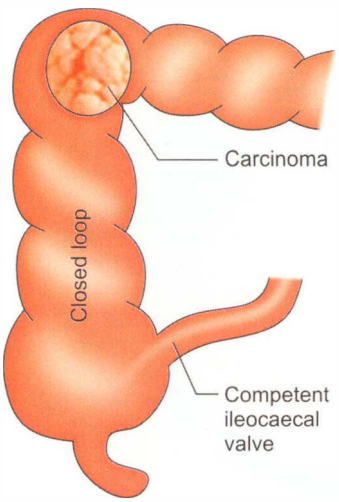


Fig. 30.2: Closed loop obstruction in 40% of patients ileocaecal valve is competent

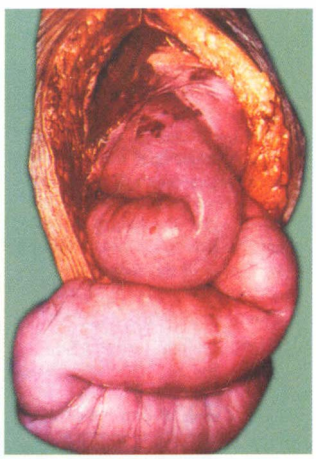


Fig. 30.3: Dilated small intestinal loops in a case of ileal obstruction

I. Depending upon the cause of obstruction

A. In the lumen of the gut

Gall stones ileus
Food bolus obstruction
Roundworm mass
Foreign body (rare)
Meconium ileus

B. In the wall of the gut

Stricture, e.g. tuberculosis
Crohn's disease
Carcinoma
Atresia
Adhesions

C. Outside the wall of the gut

Volvulus, intussusception
Congenital bands
Meckel's diverticulum with band
Obstructed hernia

II. Depending upon severity of obstruction

A. Acute obstruction: Signs and symptoms appear very early. Usually, it affects small bowel, obstructed hernia, bands.

B. Chronic obstruction (e.g. carcinoma colon) affects large bowel (colic comes first, distension later). Diverticular disease also produces chronic obstructions (Fig. 30.3).

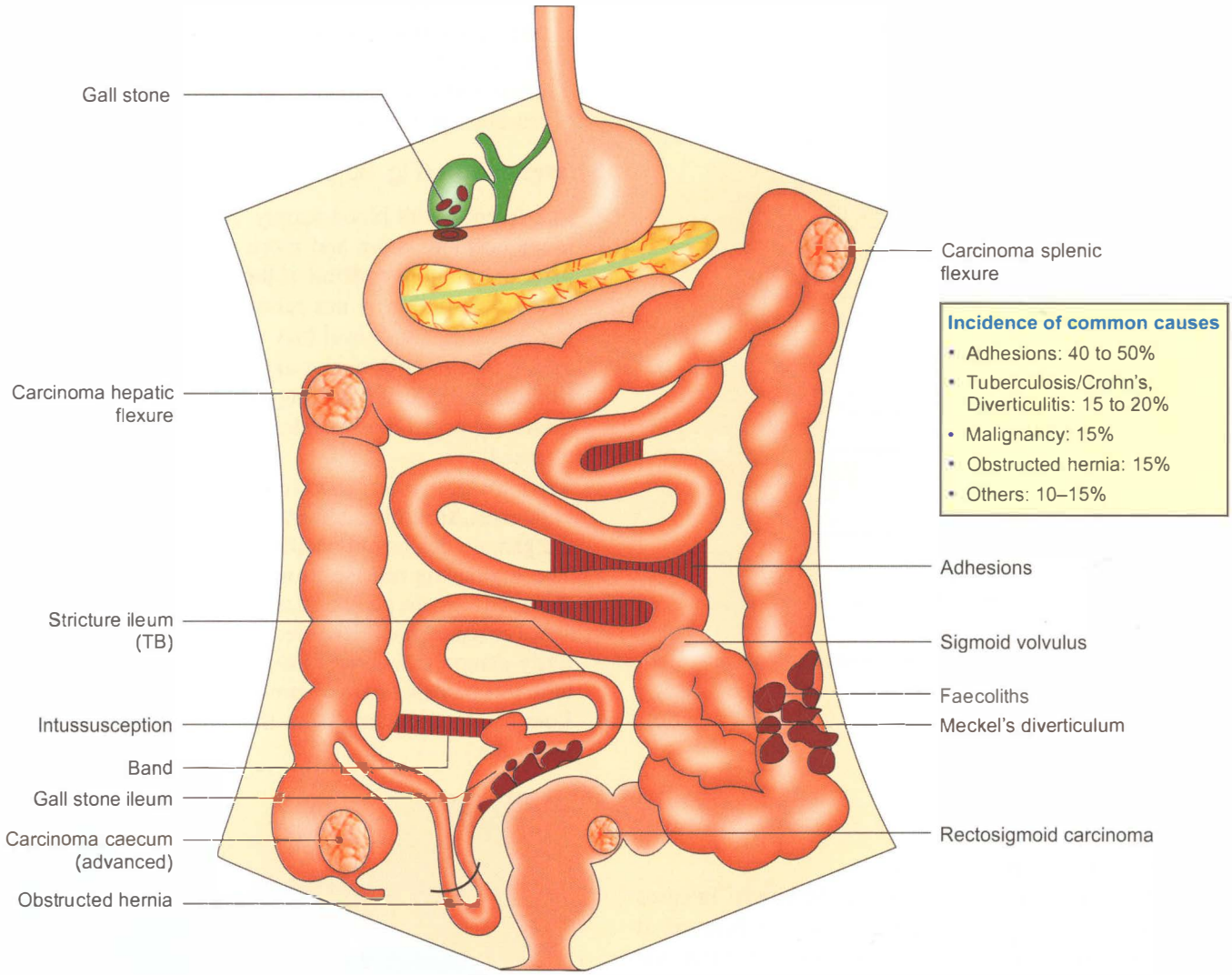
C. Acute on chronic obstruction develops in carcinoma colon, wherein an acute obstruction suddenly results due to the accumulation of faecal matter in the proximal bowel (*see* Fig. 30.4 for various causes of intestinal obstruction).

Pathophysiology (Fig. 30.5)

- As a result of obstruction, the proximal bowel undergoes hyperperistalsis which is responsible for colicky pain abdomen. The peristalsis may continue for a few days and later the intestine may be paralysed and flaccid. After 3–4 hours, distal to the obstruction, all physiological activities of the bowel are stopped. Intestine becomes contracted, pale and does not exhibit peristalsis. After a few hours, the proximal bowel gets dilated secondary to obstruction.
- The causes of distension of intestinal loop are:

A. Gaseous distension

- Swallowed air (70%). Because of colic and anxiety, the swallowed air is increased. Oxygen is absorbed and nitrogen remains as it cannot be absorbed. This results in distension.
- Diffusion of air from the blood into bowel lumen increases carbon dioxide which diffuses very rapidly.
- Gas due to bacterial activity releases H_2S, NH_3, etc.



COMMON CAUSES OF ILEAL OBSTRUCTION

- Adhesions
- Obstructed hernia
- Stricture
- Intussusception
- Ileocaecal tuberculosis
- Bands
- Worm ball—in children
- Ileal atresia—in children

COMMON CAUSES OF COLONIC OBSTRUCTION

- Carcinoma colon
- Sigmoid volvulus
- Faecal impaction
- Mesenteric ischaemia
- Hirschsprung's disease
- Anorectal malformations
- Stricture colon—rare

COMMON CAUSES OF GANGRENE

- Volvulus
- Intussusception
- Obstructed hernia
- Mesenteric vascular occlusion
- Twisting around a band
- Necrotising enterocolitis

Fig. 30.4: Differential diagnosis of intestinal obstruction—diagrammatic representation

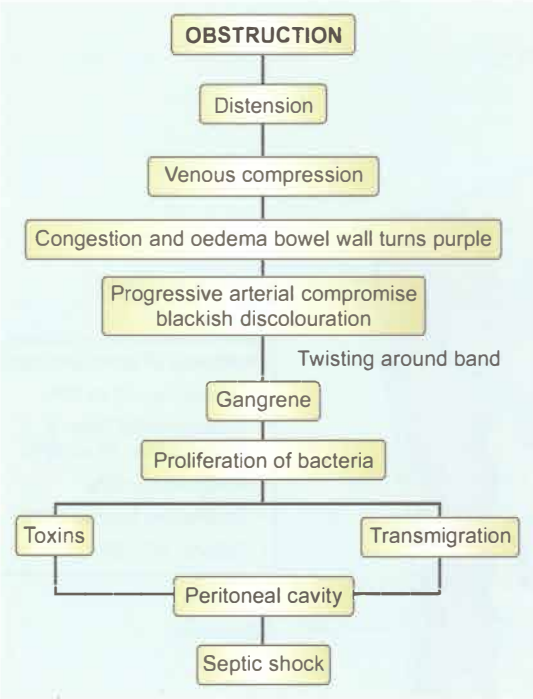


Fig. 30.5: Pathophysiology of intestinal obstruction

B. Distension due to fluids

- 1500 ml of saliva
- 2 litres of gastric juice
- 3 litres of intestinal secretions
- 1 litre of bile and pancreatic juice

Normally, all this fluid is absorbed in the bowel. In cases of intestinal obstruction, this fluid absorption is delayed. It accumulates in the intestinal loop. Excretion of water and electrolytes into the lumen is also increased.

C. Role of nitric oxide

Activated neutrophils and macrophages accumulate within the muscular layer of the bowel wall due to dilatation and inflammation of the bowel wall. This damages the secretory and motor processes by release of reactive proteolytic enzymes and cytokines. Net result is increase in **the local release of nitric oxide**, itself a potent inhibitor of smooth muscle tone. It further aggravates the intestinal dilatation.

D. Role of bacteria

- Bacterial colony count increases following obstruction resulting in stasis. From less than 10^6 in jejunum and from 10^8 in ileum, counts increase.
- Bacterial translocation can occur even in simple obstruction without strangulation. Thus, bacteria can enter into lymph nodes and into systemic circulation. Abdominal distension, hypovolaemia, renal failure and sepsis set in. In addition to these changes, diaphragm gets elevated, respiration is impaired which result in respiratory complications such as atelectasis and basal pneumonia.
- In doubtful cases of viability, if facilities are available, a test called **fluorescein** test can be done. 1000 mg of fluorescein is injected into peripheral vein and bowel is inspected under Wood light. If loops are nonviable, resection and anastomosis is done.

Strangulation (Fig. 30.6 and Key Box 30.2)

- Interference with blood supply: As the tension within the loops becomes more and more, venous congestion takes place resulting in oedema of the bowel wall.
- If the obstruction is not relieved, capillary rupture and haemorrhage into bowel may ensue. **In cases of volvulus and intussusception,** the **arterial supply gets compromised rapidly causing gangrene of bowel wall** very early. Bacterial proliferation takes place and endotoxins are released.
- **Transmigration** (translocation) of gram-negative organisms, anaerobes and gram-positive organisms through the gangrenous bowel results in peritonitis.
- The organisms release **powerful endotoxins** which are absorbed from the peritoneal surface and cause gram-negative shock or septic shock. It carries very high mortality rate (30%).
- Early gangrene without obstruction is a feature of mesenteric thrombosis or embolism.
- Loss of blood volume is an important feature of massive gangrene.

Fig. 30.6: Gangrene of the intestine due to bands

KEY BOX 30.2

FACTORS PREDISPOSING ISCHAEMIA

- Volvulus
- Mesenteric ischaemia
- Necrotising enterocolitis
- Intussusception
- Progressive distension
- Extrinsic compression by adhesions, bands, etc.

Fig. 30.7: Faeculent vomiting is pathognomonic of terminal ileal obstruction

Clinical features (Key Box 30.3)

1. **Pain abdomen:** Central abdominal pain is a feature of small intestinal obstruction and peripheral pain is a feature of large intestinal obstruction. The pain is colicky in nature, lasts for 5–10 minutes and is intermittent. On pressure, it decreases.
2. **Vomiting** is due to reverse peristalsis. Vomitus consists of stomach contents initially, then bile, followed by faeculent matter. **Faeculent is not faecal matter** but **terminal ileal contents which undergo bacterial degradation** and **fermentation** resulting in the smell of faecal matter. Vomiting of altered blood indicates haemorrhage and gangrene. Frequent vomiting reflects jejunal obstruction (Fig. 30.7 and Table 30.1).

PEARLS OF WISDOM

Vomiting of faeculent contents indicates terminal ileal obstruction.

3. **Distension of the abdomen:** It may be central abdominal distension as seen in ileal obstruction, peripheral abdominal as in large bowel obstruction, or localised to one or two quadrants as in sigmoid volvulus.
4. **Constipation** occurs because the distal bowel does not move. Constipation to faeces and flatus is called obstipation. Exceptions are given in Key Box 30.4.

Signs

1. **General signs of dehydration** such as dry skin, dry tongue, sunken eyes, feeble pulse, low urinary output are seen. Dehydration occurs due to persistent vomiting and sequestration of fluid and electrolytes. Hypokalaemia is an important finding.
2. **Abdominal findings**
   - Distension, tympanitic note on percussion
   - Step ladder peristalsis is seen in terminal ileal obstruction. Right to left colonic peristalsis is seen in left-sided colonic obstruction, large bowel obstruction.
   - On auscultation—loud, noisy intestinal sounds are heard. They are called **borborygmi**.
   - Hernial orifices have to be examined, especially for a femoral hernia in females.

Signs of strangulation

- It should be suspected when features of obstruction are present along with features of shock.

KEY BOX 30.3

CARDINAL FEATURES OF INTESTINAL OBSTRUCTION

- Colicky abdominal pain
- Abdominal distension
- Vomiting
- Absolute constipation

KEY BOX 30.4

INTESTINAL OBSTRUCTION WITH DIARRHOEA

- Faecal impaction : Page 731 (Fig. 30.4)
- Richter's hernia : Page 858
- Gall stone ileus : Page 747
- Mesenteric vascular occlusion : Page 753

Table 30.1 Comparison of clinical features at different levels of intestinal obstruction

	High (jejunum)	Distal (ileum)	Low (colon)
• Vomiting	Frequent, bilious	Moderate bilious, faeculent	Late vomiting, faeculent
• Distension	No	Moderate distension	Marked distension
• Pain	Intermittent, not crescendo type	Intermittent, crescendo type, colicky	Variable pain, not classical crescendo
• Constipation	Not initially	Not initially	Initially present
• Peristalsis	Not seen	Step ladder peristalsis	Right to left peristalsis may be seen

Fig. 30.8: Dilated intestinal loops

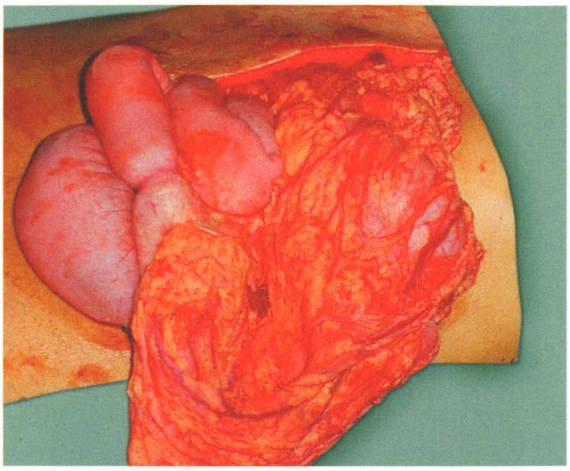


Fig. 30.9: Perforation due to obstruction

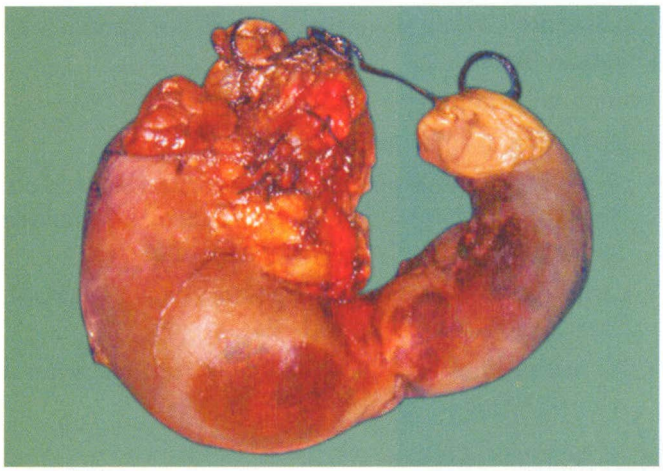


Fig. 30.10: Gangrene due to volvulus

Fig. 30.11: Step ladder peristalsis

- Features of **septic shock**—fever, hypothermia, renal failure, respiratory failure (Key Box 30.5).
- **Rebound tenderness:** It is called **Blumberg's sign**. It is a classical sign of peritonitis.
- Guarding and rigidity of the abdominal wall.
- **Absent bowel sounds** because rest of the bowel loops undergo paralytic ileus.
- **Sudden symptoms**—spasmodic pain (due to peristalsis) and continuous pain suggest strangulation (Fig. 30.11).
- Features of strangulation and perforation occur quickly in cases of closed loop obstruction (Figs 30.8 to 30.11 and Key Box 30.6).

KEY BOX 30.5

FEATURES OF STRANGULATION

- Tachycardia
- Tenderness
- Temperature—fever
- Acidosis

KEY BOX 30.6

CLOSED LOOP OBSTRUCTION

- This occurs when the bowel is obstructed at both proximal and distal points.
- Proximal bowel is not distended as much in this condition
- Gangrene and perforation can occur fast
- Retrograde thrombosis of mesenteric vein, can result in distension of the bowel.
- A few examples of closed loop obstruction include sigmoid volvulus, strangulated hernia, carcinoma right colon.

Rectal examination

- In small bowel obstruction, **rectum is empty** and is often ballooned out.
- Carcinomatous growth with or without stools can be felt.
- The finger may be stained with blood.

- The small intestine is considered dilated if loops of bowel measure **more than 3 cm** in diameter. Measurements for the large bowel vary among different anatomic segments, with a relative threshold of **9 cm in diameter for the proximal colon** and **5 cm for the sigmoid colon.**

INVESTIGATIONS

- **Complete blood picture:** Low Hb% indicates underlying malignancy. Increased total WBC count indicates infection and sepsis (perforation).
- **Electrolytes:** Most of the electrolytes are low in cases of intestinal obstruction and require correction preoperatively. Strangulation may be associated with deranged potassium, amylase or lactic dehydrogenase.
- **Plain X-ray abdomen** in the erect position may show multiple gas fluid levels. Gas levels appear earlier than fluid level. Normally, two insignificant fluid levels can be present, one in the terminal ileum and one in the first part of the duodenum (Key Box 30.7). **Supine films indicate the distal limit of obstruction** (Figs 30.12 to 30.16).

PEARLS OF WISDOM

Enteroclysis is rarely performed in acute intestinal obstruction but it has greater sensitivity in the detection of partial small bowel obstruction (Fig. 30.17).

- **Ultrasound/CT scan**—*See* Fig. 30.18, Key Boxes 30.8 and 30.9

KEY BOX 30.7

PLAIN X-RAY FINDINGS UPRIGHT AND SUPINE

- First get supine films. They indicate distal limit of obstruction. Erect films are asked if any doubt exists about obstruction.
- Jejunum is characterised by regularly placed mucosal folds called **valvulae conniventes** (Fig. 30.14) placed opposite to each other **(Herring bone pattern)**. They are produced by **valves of Kerckring.**
- Large bowel is characterised by **haustrations** (Fig. 30.15): Incomplete, large mucosal folds, not placed opposite to each other.
- **Caecum has no haustrations.** It appears as a round gas shadow in the right iliac fossa.
- Ileum has no characters—**characterless loop of Wangensteen**.
- Plain X-ray may demonstrate gall stone ileus or foreign body.
- Gas is absent in the small bowel as in mesenteric vascular ischaemia.
- Sigmoid volvulus appears as a large dilated loop—inverted 'U' shape.

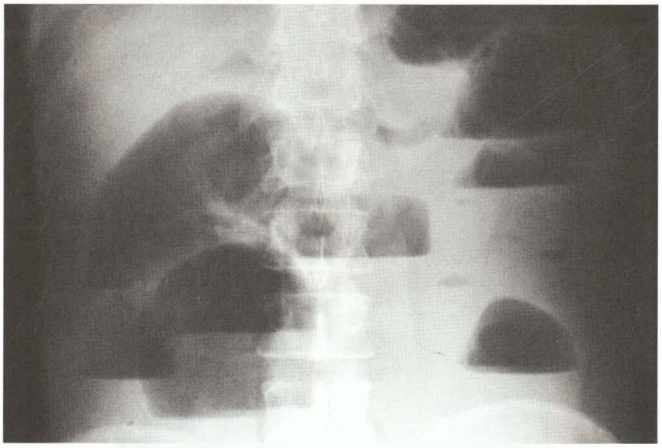


Fig. 30.12: Multiple gas fluid levels—plain X-ray erect abdomen

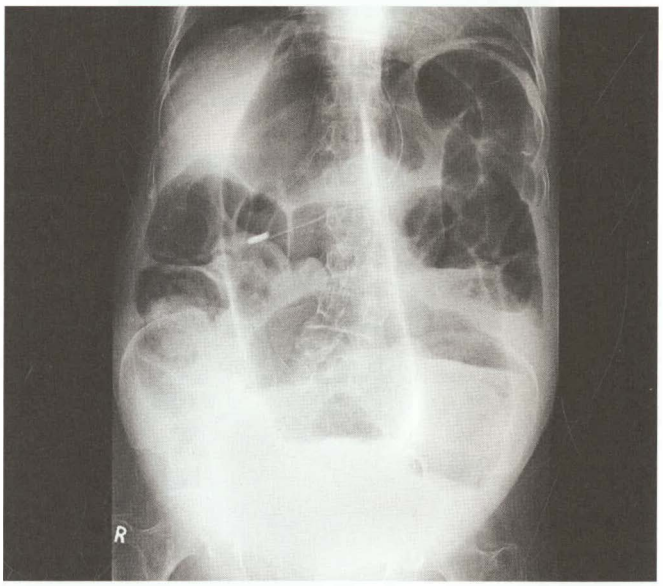


Fig. 30.13: Multiple gas fluid levels seen with a large dilated loop in the centre

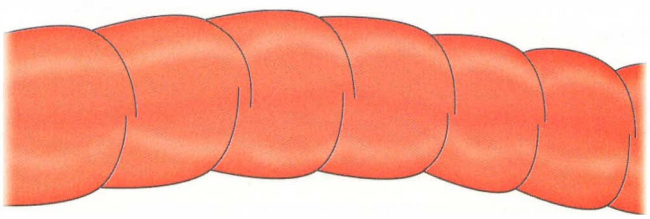


Fig. 30.14: Valvulae conniventes

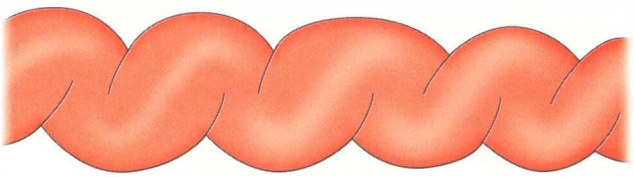


Fig. 30.15: Haustrations

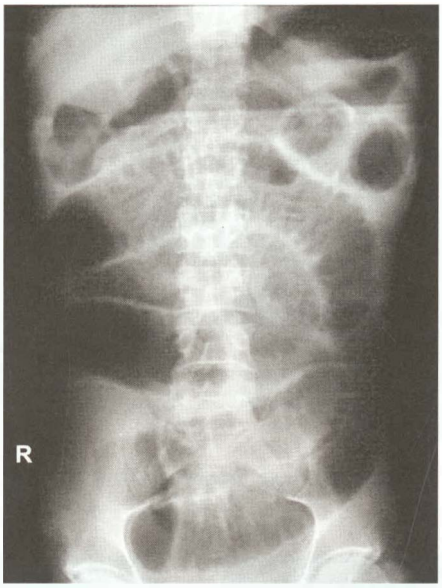


Fig. 30.16: Plain X-ray abdomen supine—distended intestines. Supine X-ray will indicate distal limit of obstruction

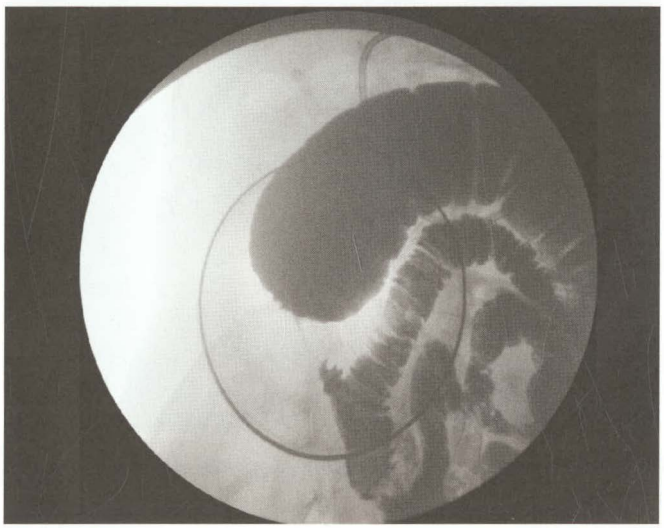


Fig. 30.17: Jejunal stricture due to tuberculosis—enteroclysis picture

KEY BOX 30.8

ULTRASOUND

- It is not the investigation of choice or it may not be required

However, a good sonologist can diagnose:

a. Dilated loops of intestine
b. Presence of fluid in the abdomen
c. Gall stone ileus
d. Infarcted/ischaemic bowel/gas in the portal vein or intrahepatic gas.
e. Intussusception and can assess vascularity with the help of duplex scan.
f. It can also rule out other causes

Being a noninvasive investigation, it has more benefits

KEY BOX 30.9

CT SCAN IN INTESTINAL OBSTRUCTION

- Can detect dilated intestines proximally and collapsed bowel distally.
- If bowel wall is thick and air is present (**pneumatosis**), strangulation is likely.
- It can detect portal venous gas (suggesting gangrene)
- CT can detect **mass lesions**—carcinoma sigmoid, caecum or ileocaecal mass (TB).
- CT has low sensitivity in detecting low grade or partial small bowel obstruction. Sensitivity increases in total obstruction.

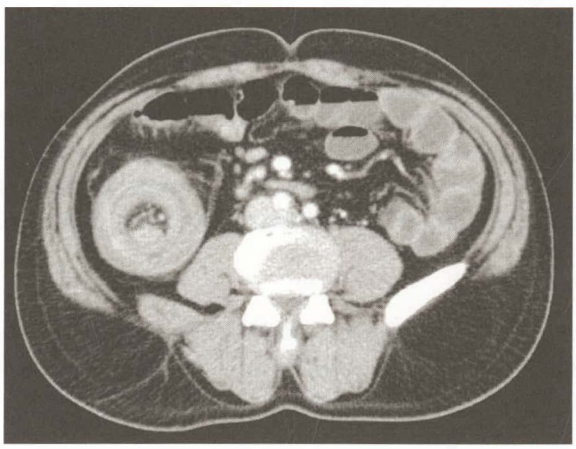


Fig. 30.18: Ileocolic intussusception—observe 'target sign' on the right side

MANAGEMENT

Preoperative preparation includes correction of dehydration, electrolytes and broad spectrum antibiotics. Principles in the management of intestinal obstruction are as follows:

A. Aspiration with Ryle's tube. This is the most important step in the management of intestinal obstruction. It helps in decreasing the distension and also prevents vomiting. This will help in preventing respiratory complications, such as aspiration following general anaesthesia.

B. Bowel care: No purgatives because purgation can cause perforation.

C. Charts: Temperature, pulse, respiration and intake-output chart. In cases of conservative management such as obstruction due to adhesions, change in temperature and increasing pulse rate suggests perforation or gangrene. These cases have to be explored immediately.

D. Drugs to cover gram-positive, gram-negative and anaerobic organisms.

E. Exploratory laparotomy is done and depending upon the findings, obstruction is treated. A few examples are given in Key Box 30.10 and Fig. 30.19.

F. Fluids should be given before, during and after surgery. It forms the most important treatment of intestinal obstruction.

KEY BOX 30.10

PRINCIPLES OF EXPLORATORY LAPAROTOMY

- Ideally done within 6–8 hours
- Long midline incision
- Resection of gangrene and anastomosis
- Adhesion—release
- Bands—divide
- Gall stone ileus—remove the stone
- Volvulus—untwist or resection
- Obstructed hernia—reduce
- Gangrene—resect
- Stricture—resection or stricturoplasty
- Advanced malignancy—bypass

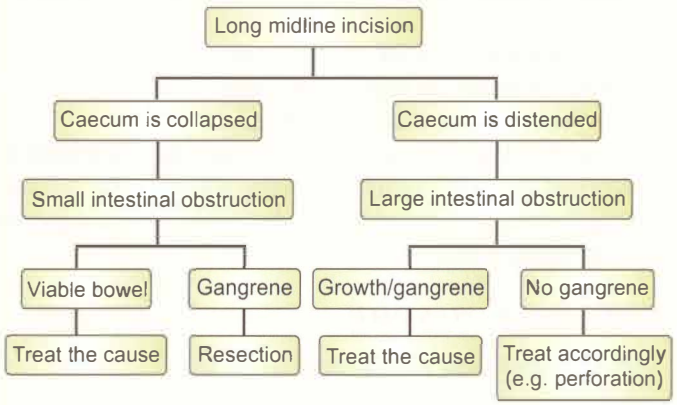


Fig. 30.19: Principles of management of intestinal obstruction

APPROACH TO THE MANAGEMENT OF INTESTINAL OBSTRUCTION

Ask the following questions to yourself and proceed.

1. What is the probable cause of obstruction?
2. Is it small bowel obstruction at laparotomy?
3. Is it large bowel obstruction at laparotomy?
4. Is it simple obstruction?
5. Is it strangulation?
6. Is it some kind of a surprise or a difficult case?
7. Can I manage conservatively?

1. Probable cause of obstruction

- A previous laparotomy scar may indicate that it could be an adhesive obstruction (most common).
- An obvious obstructed hernia (inguinal or femoral) can be managed with inguinal approach.
- An elderly man, hypertensive and atherosclerotic, with features of blood in the stools and acute abdominal pain may be having superior mesenteric ischaemia.
- A constipated, elderly man in poor health, with acute or chronic obstruction may be having carcinoma of the colon.

2. Diagnosis of small bowel obstruction at laparotomy

- Caecum is collapsed
- Dilated loops of small intestine are present.
- A stricture or a mass lesion may be obvious at laparotomy.

3. Diagnosis of simple obstruction

- It is done when bowel is not gangrenous.
- In doubtful cases, because of long-standing ischaemia, wrapping the bowel with warm and moist pack and administration of pure oxygen may help the bowel to recover from ischaemia.

4. Diagnosis of large bowel obstruction at laparotomy

- Caecum is distended.
- A growth may be palpable and obvious in the transverse colon or in the hidden colon, i.e. splenic flexure.
- It is very important to examine the entire colon (synchronous carcinoma is more common).

5. Diagnosis of strangulation

(Fig. 30.20 and Key Box 30.11)

- Black, dark, foul-smelling bowel is seen as soon as laparotomy is done.
- Peritoneal fluid contains blood-stained fluid.
- Precautions must be taken not to contaminate peritoneal cavity when gangrenous segment is removed.

Do not hesitate to take the help of senior experienced surgeons in treating an uncommon situation such as massive ischaemia and gangrene of small bowel and colon (due to mesenteric vascular occlusion), synchronous carcinoma and ileosigmoid knotting, etc.

KEY BOX 30.11

VIABLE BOWEL—FEATURES

- Normal peristalsis
- Normal peritoneal sheen is present
- Normal pulsations are visible or felt at the mesentery
- Normal pink colour is present

Fig. 30.20: Viable and nonviable bowel

6. It is a surprise

- Surprises are well known in intestinal obstruction. Congenital bands, foreign bodies, internal herniation, lymphomatous strictures are a few examples.
- The detailed management of individual cases is discussed below.

7. Can I manage conservatively?

- In these cases, a long intestinal tube called Miller-Abbott tube can be passed to decompress intestines (Key Box 30.12).

KEY BOX 30.12

INTESTINAL OBSTRUCTION
CONSERVATIVE TREATMENT

1. **Partial small bowel obstruction** mostly due to adhesion: Wait for 48 to 72 hours. They may show improvement. If not, surgery is required.
2. **Early postoperative obstruction:** It rarely progresses to strangulation. Hence, nonoperative management can be extended to many days (3–7) provided there is no evidence of peritonitis.
3. **Intestinal obstruction in Crohn's disease:** Aim in Crohn's disease is to 'preserve' bowel as it may respond to medications.
4. **Carcinomatosis:** Disseminated malignancy with obstruction. The aim is nonoperative treatment as nothing much can be achieved with laparotomy.

DIFFERENTIAL DIAGNOSIS OF INTESTINAL OBSTRUCTION

VOLVULUS OF THE SIGMOID COLON

- Common in North India (Punjab), Eastern Europe, Uganda.
- In certain parts of India as mentioned above, it is one of the common surgical emergencies in elderly population (Key Box 30.13).

KEY BOX 30.13

CERTAIN FACTS ABOUT SIGMOID VOLVULUS

- More common in males
- 2/3 of the cases are sigmoid volvulus and 1/3 are caecal volvulus.
- Common in middle age and > 60 years
- It consists about 10 to 15% of cases of intestinal obstruction in India.
- More common in rural population
- It is not an uncommon event during pregnancy

Precipitating factors (Fig. 30.21)

1. Long mesentery of the pelvic colon
2. Narrow attachment at the base

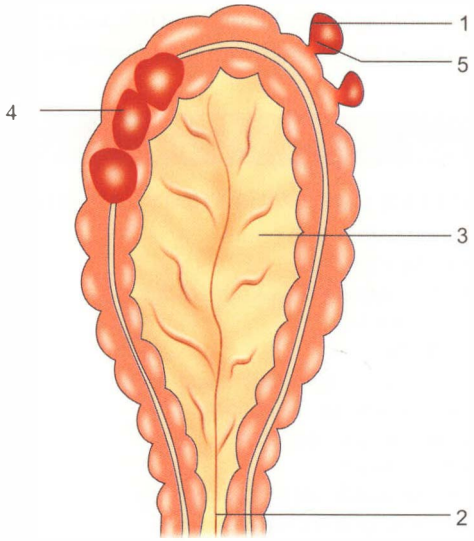


Fig. 30.21: Causes of sigmoid volvulus (see text for numbers)

3. Long, redundant, pendulous sigmoid
4. Loaded colon due to high residue diet
5. Diverticulitis with a band, or adhesions

Sigmoid volvulus is a definite occurrence in mentally disturbed patients, hypothyroidism, Parkinson's disease, multiple sclerosis, etc. probably due to severe constipation due to medications.

PEARLS OF WISDOM

Ogilvie's syndrome precipitates volvulus.

Clinical features

1. **Acute sigmoid volvulus (fulminant)** presents as intestinal obstruction. It starts usually after straining at stools. Volvulus is usually in anticlockwise direction and after one and a half turns, the entire loop becomes gangrenous.

   Enormous distension of the abdomen takes place, which gives a tympanitic note all over the abdomen. It is due to **diffusion of CO_2** (Fig. 30.22). Due to gross distension,

Fig. 30.22: Uneven distension due to sigmoid volvulus

severe hypovolaemic shock develops within 6–8 hours of volvulus. Gangrene sets in, which gives rise to features of strangulation. A dilated loop can be seen and felt. Features of peritonitis are seen within 1–2 days. Per rectal examination shows rectum to be empty.

PEARLS OF WISDOM

Distended tympanitic drum like abdomen—sigmoid volvulus.

2. **Chronic (indolent) recurrent sigmoid volvulus:** It occurs due to partial twisting and untwisting of the bowel. Elderly patients present with recurrent lower abdominal pain on the left side and distension of the abdomen which is relieved on passing large amount of flatus.

Diagnosis

- Plain X-ray abdomen erect shows a hugely dilated sigmoid loop which is described as **'bent inner tube sign'**. The dilated loop may be visible on the right side, centre and to the left of abdomen, having two fluid levels, one on right side and one on left side. This is also described as **'omega sign'** (Figs 30.23 and 30.24).
- **Contrast enema:** As the barium enters the rectum, it tapers into the sigmoid colon—**Bird's beak sign**.

Treatment

I. Nonoperative

- A successful passage of flatus tube or sigmoidoscope up to 25–30 cm results in release of a large amount of flatus and fluid and obstruction is relieved. If obstruction is completely relieved or if there is no gangrene and the general condition of the patient improves, an elective resection is done after 7 days. If resistance is found while passing flatus tube, instill barium for guidance.

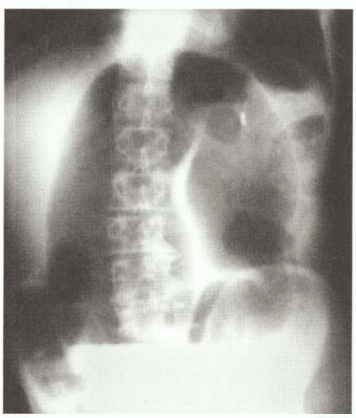


Fig. 30.23: Plain X-ray abdomen showing distended sigmoid loop

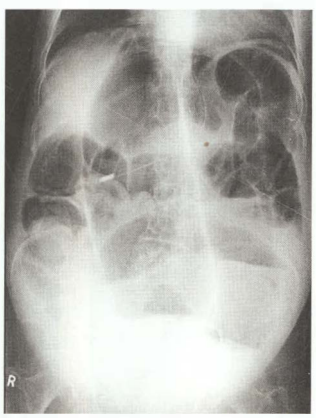


Fig. 30.24: An 84-year-old patient with sigmoid volvulus, managed by sigmoidoscopic decompression

II. Operative treatment

1. **Single-stage resection:** This can be done, provided general condition of the patient is good. If the loop is gangrenous, resection followed by end to end anastomosis is done, after giving 'on table' lavage using saline washes till the contents of the colon are clear. Sigmoid colon is hugely dilated (Figs 30.25 and 30.26).
2. **Hartmann's procedure:** If the loop is gangrenous and proximal bowel is loaded with faecal matter, resection of the sigmoid colon is done. Proximal descending colon is brought out as an end colostomy and rectum is closed (Hartmann's procedure). After 6 weeks, colorectal anastomosis is done.
3. **Sigmoidopexy:** If the loop is not gangrenous, untwist the sigmoid loop and fix the sigmoid to the posterior abdominal wall (sigmoidopexy). If the mesentery is long, it can be made short by plication.
4. **Exteriorisation:** Paul-Mickulicz procedure is done when general condition of the patient is poor as in elderly patients, in severely dehydrated patient with impending septicaemia. In such cases, the gangrenous loop is brought outside and resected, with a proximal colostomy and a distal mucous fistula (Fig. 30.27).

Fig. 30.25: Sigmoid colon at surgery—huge distension results in severe hypovolaemic shock

Fig. 30.26: Distended sigmoid with venous congestion

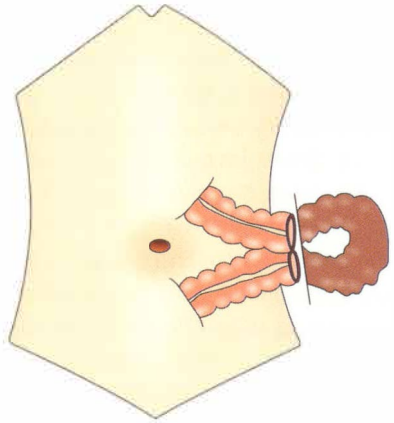


Fig. 30.27: Paul-Mickulicz procedure

CAECAL VOLVULUS AND BASCULE

- It is a rare cause of intestinal obstruction (Figs 30.28 to 30.32 and Key Boxes 30.14 and 30.15).
- Presumably, it is more likely to occur following any surgical procedure which might require some degree of medial visceral rotation or disruption of the fusion plane between the caecum or ascending colon with the lateral peritoneum, providing sufficient mobility allowing caecal volvulisation to occur.
- A constant feature of **caecal bascule** is the presence of a constricting band across the ascending colon. This may be found at laparotomy.
- In a plain X-ray abdomen, caecum produces round shadow in the centre of the abdomen.
- Resection is the ideal treatment.

PEARLS OF WISDOM

If loop is gangrenous, do not derotate—clamp the mesentery first, then detorsion can be done to avoid reperfusion injury.

KEY BOX 30.14

COMPARISON OF CAECAL VOLVULUS AND SIGMOID VOLVULUS

Caecal volvulus	Sigmoid volvulus
• Rare	• Common
• Clockwise twist	• Anticlockwise
• Mobile caecum	• Long mesentery is the cause
• Middle-aged	• Elderly, debilitated
• Kidney-shaped gas shadow with single fluid level on the left side.	• Omega sign or coffee bean-shaped. Two fluid levels can be found.
• Treated only by surgery	• Nonoperative treatment should be attempted in all cases, provided there is no ischaemia.

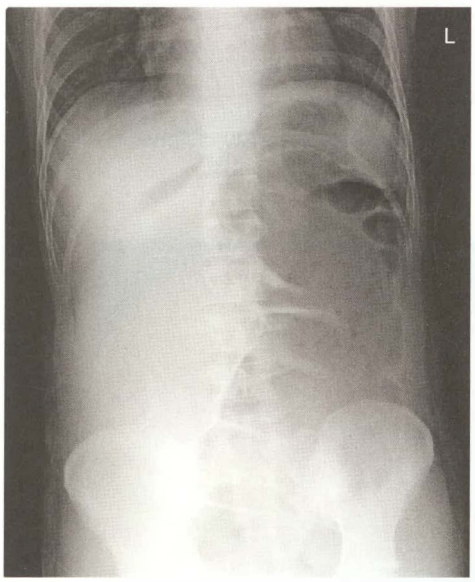


Fig. 30.28: Supine X-ray findings in caecal volvulus

Fig. 30.29: Erect X-ray showing distended loop. Caecum was palpable in the left hypochondrium

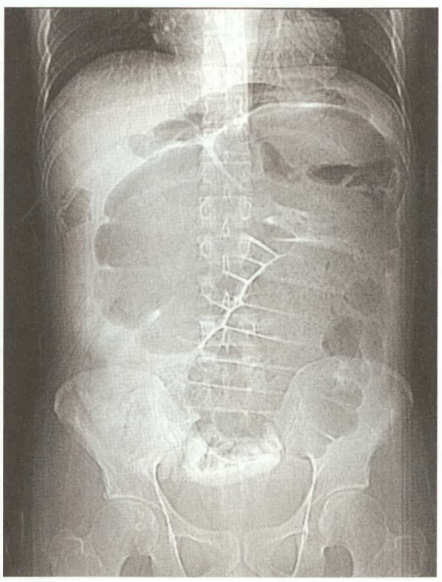


Fig. 30.30: CT X-ray showing dilated coils of bowel

Fig. 30.31: Distended caecum at surgery
(*Courtesy:* Dr Basavaraj Patil, Dr Hartimath, Dr Rasheed, Dr Nikil, Department of Surgery, KMC, Manipal)

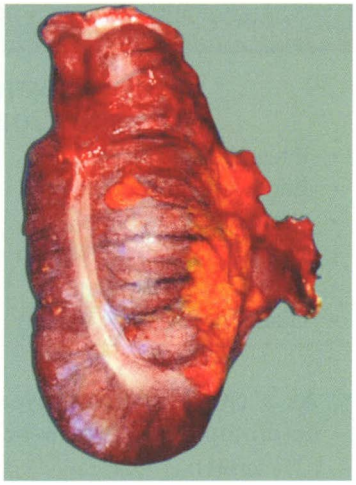


Fig. 30.32: Caecal volvulus with gangrene

KEY BOX 30.15

CAECAL BASCULE

- Caecum folds anteromedial to the ascending colon, with production of a flap-valve occlusion at the site of flexion.
- Caecum will be markedly distended and will be found in the centre of the abdomen.
- Occasionally, it is associated with malrotation of the gut
- There will be a constricting band across the ascending colon.
- It is also argued that caecal bascule can also be due to lack of fixation of large bowel. Resection is ideal even though fixation (caecopexy) is also another alternative.
- Bascule is a French term for see-saw and balance

MECKEL'S DIVERTICULUM WITH A BAND

It is a congenital diverticulum which occurs due to persistent intestinal end of vitellointestinal duct. **Being congenital, it has all the layers of the bowel**. Hence, a true diverticulum.

Anomalies of vitellointestinal duct (Fig. 30.33)

A. Fibrous band results when entire duct is obliterated and bowel can twist around the band, resulting in volvulus.

B. Persistent intestinal end: Meckel's diverticulum (Key Box 30.16).

C. Meckel's diverticulum with the band attached to the umbilicus can give rise to intestinal obstruction.

D. Umbilical fistula results when entire duct is patent. Even though it is connected to the terminal ileum, the opening is very small. The discharge is rarely faecal. Often, it is the mucus secreted from the lining of the duct **(omphaloenteric fistula)**.

E. Umbilical sinus results due to persistent umbilical end discharging mucus. Slowly umbilical adenoma occurs and epithelial lining of the sinus gets everted.

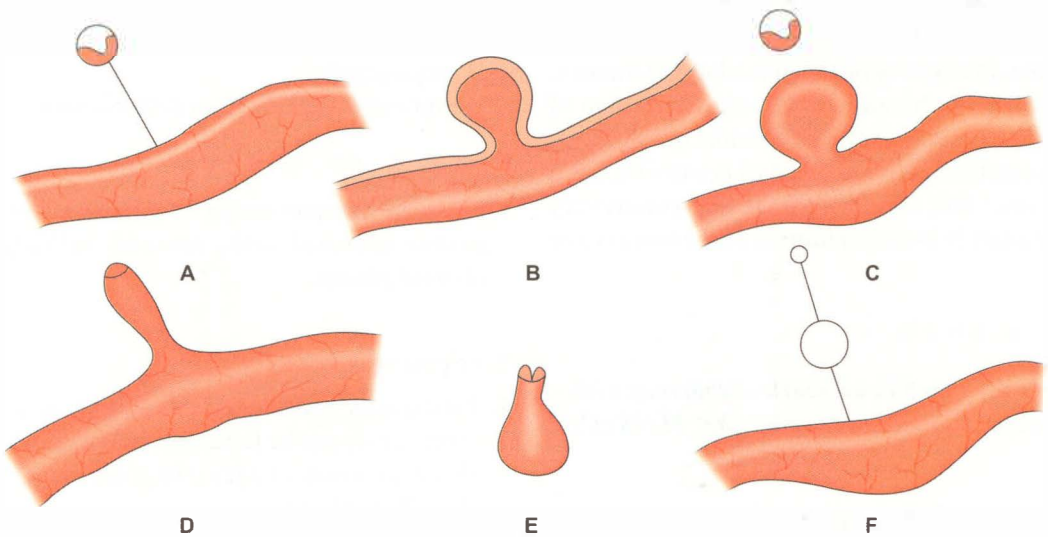
F. Intra-abdominal cyst results when both ends are obliterated. The central portion of the duct persists and secretes mucus. This is very, very rare.

KEY BOX 30.16

MECKEL'S[1] DIVERTICULUM AND OTHER ASSOCIATED ANOMALIES

- Angiodysplasia of the caecum
- Anorectal atresia, atresia of the oesophagus

[1]The most common congenital anomaly of small intestine



Figs 30.33A to F: Anomalies of vitellointestinal duct (A to F *see* text for details)

MECKEL'S DIVERTICULUM

- It is present in 2% of the cases, 2 inches long, 2 feet away from ileocaecal region in the antimesenteric border (ileal duplication can occur in the mesenteric border). It is two times more common in females.
- Symptomatic cases are below 2 years of age.
- In 12% of the patients, heterotopic gastric tissue is found which can produce peptic ulceration. In a few other patients, it can contain pancreatic and colonic tissue.

Rule of 2 for Meckel's diverticulum

- Incidence: 2%
- Location: 2 feet proximal to ileoceacal junction
- Length: 2 inches long
- Ectopic tissue: 2 types—gastric and pancreatic
- Presentation: 2 years or below 2 years is the most common age
- Male: female ratio—1:2

Clinical presentation

- **Massive bleeding per rectum:** In the form of melaena, it is not uncommon. In many other patients, mild chronic bleeding can result in anaemia.
- **Acute Meckel's diverticulitis:** Factors which precipitate diverticulitis are:
  - Peptic ulceration due to ectopic gastric mucosa of the diverticulum
  - Ingested foreign material, e.g. stalk of vegetable, seeds, fish or chicken bones
  - Faecolith (not common in ileum), tumours, worms causing stasis and bacterial infection
  - Inflammation and ischaemia caused by torsion due to an associated band is called mesodiverticular band
  - Association with acute appendicitis

  Such cases can perforate very often. It is impossible to differentiate it from ruptured appendix. This is treated by laparotomy and resection of diverticulum along with adjacent intestine. In majority of appendicular perforations, local abscess will occur because of retrocaecal position of the appendix (70%). However, perforation of Meckel's diverticulum, even though a rare cause of peritonitis, has a high mortality rate. This is because infection spreads very fast as **diverticulum is intraperitoneal and contents are faeculent**.

PEARLS OF WISDOM

Older child, deformed umbilicus, scarless abdomen with intestinal obstruction—cause may be Meckel's diverticulum.

- As a cause of **intestinal obstruction**, when it is associated with a band or due to volvulus. It is the most common presentation in adults (Key Box 30.17).

KEY BOX 30.17

MECKEL'S DIVERTICULUM AND INTESTINAL OBSTRUCTION

- Intussusception
- Band
- Volvulus due to band
- Internal herniation beneath mesodiverticular band
- Diverticulitis with band
- Littre's hernia

- **As a cause of intussusception:** Here also, inflamed heterotopic tissue can be found in the diverticulum (2% cases).
- **Pain** can occur due to chronic peptic ulceration.
- **Neoplasm:** Carcinoids and GIST are more common in Meckel's diverticulum than elsewhere in the small intestine although presence of Meckel's diverticulum itself is rare.

PEARLS OF WISDOM

Hernia of Littre is a hernial sac containing Meckel's diverticulum (Key Box 30.17).

Investigations

1. No investigation can prove diagnosis of Meckel's diverticulum. Small bowel enema may demonstrate the diverticulum if opening is wide (fluoroscopy is more ideal).
2. **^{99m}Tc-labelled pertechnetate** given IV, may localise the heterotopic gastric mucosa in the Meckel's diverticulum, in about 90% of patients.
   - This radionuclide is taken up by **mucin-secreting cells** and **parietal cells** and it is secreted immediately. Thus, if ^{99m}Tc appears in the stomach as well as in other part of bowel, it indicates functioning heterotopic tissue.
   - Even when bleeding is at a rate of 0.1 ml/minute, it can detect Meckel's diverticulum. Hence, it is superior to angiography.
   - Very useful in children with bleeding.

PEARLS OF WISDOM

The pertechnetate anion ^{99m}Tc is selectively taken up by gastric mucosal cells, thyroid, salivary glands and choroid plexus.

Treatment (Fig. 30.34)

1. Incidentally found Meckel's during laparotomy for some other causes can be left alone, provided it has a wide mouth. However, a note of it must be made in the operation register (Key Box 30.18).
2. Meckel's diverticulum with bleeding, band, perforation and narrow mouth is treated by removal of diverticulum with

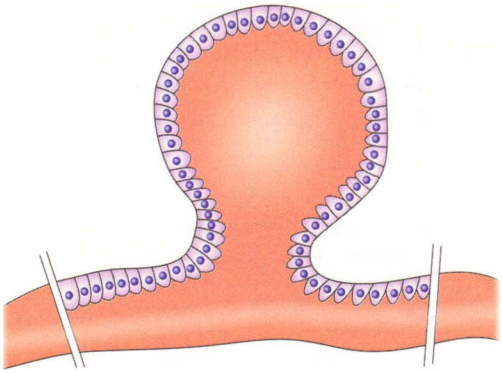


Fig. 30.34: Meckel's diverticulectomy should include normal intestine also

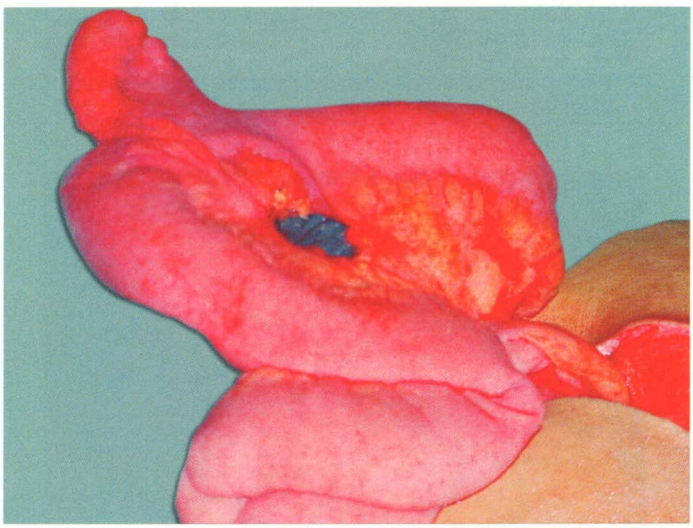


Fig. 30.35: Inflamed Meckel's diverticulum

adjacent intestine because the gastric tissue may often line the intestine also (Figs 30.35 and 30.36).

KEY BOX 30.18

INDICATIONS FOR REMOVAL OF INCIDENTALLY FOUND MECKEL'S DIVERTICULUM

- Children under 2 years of age
- Meckel's with a band
- Meckel's with adhesions
- Meckel's with narrow base
- Long Meckel's diverticulum

ADHESIONS AND BANDS

ADHESIONS

Introduction: Intra-abdominal adhesions develop after abdominal surgery as part of the normal healing processes that occur after damage to the peritoneum. The early balance between fibrin deposition and degradation seems to be the critical factor in adhesion formation. They also cause

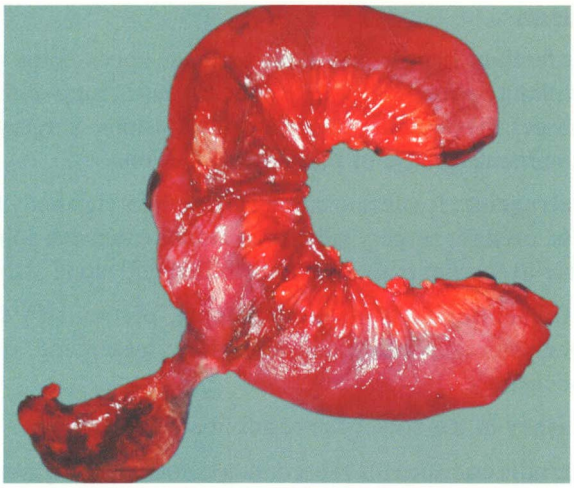


Fig. 30.36A: Resected Meckel's diverticulum with normal intestine (*Courtesy:* Dr Pramod K, Dr Deviprasad Shetty, Kasturba Hospital, Manipal)

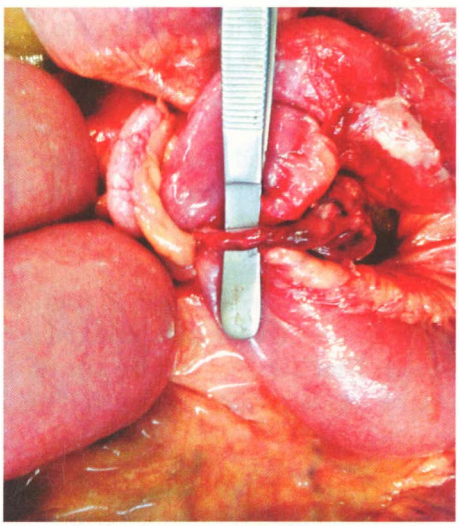


Fig. 30.36B: Meckel's diverticulum with band and obstruction

significant morbidity, including adhesive small bowel obstruction, infertility and increased difficulty with reoperative surgery. Thus, high chances of causing intestinal fistula after re-exploration.

Definition: Peritoneal adhesions can be defined as abnormal fibrous bands between organs or tissues or both in the abdominal cavity that are normally separated.

They are common causes of intestinal obstruction in the western world, Chinese population of Malaysia, etc. In India, adhesions and obstructed hernia are the two common causes in adults.

PEARLS OF WISDOM

It is important to realise that secondary infertility in women and ectopic gestation can occurs due to adhesions.

Causes

1. **Infection:** Laparotomy done for acute appendicitis with or without perforation, perforation peritonitis, intra-abdominal abscess have higher incidence of adhesions. Surgery is the commonest cause of peritoneal adhesions.
2. **Iatrogenic:** It refers to talc, silk thread, foreign body (mop), etc. used for surgery which can induce extensive adhesions due to foreign body reaction (spilled gall stone also).
3. **Ischaemia:** Lack of blood supply, particularly venous occlusion can cause adhesions, e.g. mesenteric vascular occlusion.
4. **Injury** to the bowel can result in adhesions.
5. **Irradiation** enteritis is becoming common due to irradiation for carcinoma of the cervix.

   Remember the important **5 Is**.

Pathogenesis

Ischaemia and irritation of the intestines are the chief factors responsible for adhesions (Key Box 30.19).

Types (Fig. 30.37)

1. **Fibrinous adhesions** (bread and butter adhesions): They are the causes of early postoperative obstruction, which settles down within 3–5 days. Majority of them disappear in due course of time.
2. **Fibrous adhesions:** If the infection is continuing or if foreign body is present, the fibrinous material is converted to fibrous band. They also occur at the site of ischaemia. They will cause late intestinal obstruction (Figs 30.38 and 30.39).
3. **Tuberculous adhesions** are dense adhesions that result in matting of intestinal coils. Separating them at laparotomy is extremely difficult (Fig. 30.40).
   - Congenital band is the 4th type of band (not due to adhesion).

PEARLS OF WISDOM

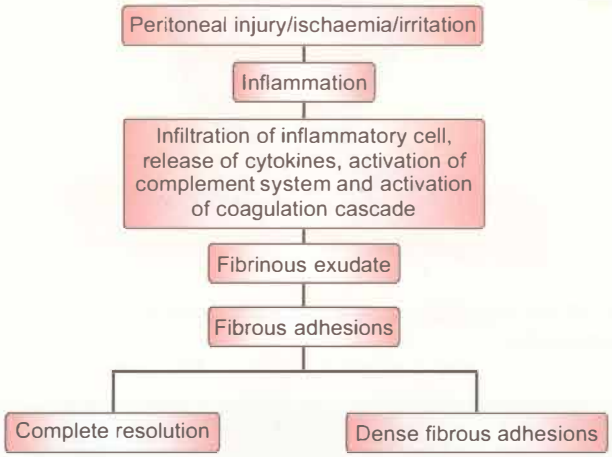
A congenital band, omental band, a string band of bacterial peritonitis or a fibrous band of tuberculosis is responsible for obstruction (Figs 30.41, 30.42 and Key Box 30.20).

Clinical features

- Recurrent abdominal pain, vomiting and distension are the typical features. Often the attacks are mild and self limiting. But persisting symptoms require monitoring and treatment.
- There may be peristalsis as in cases of terminal ileum.
- Gangrene is not common in cases of adhesive obstruction (Figs 30.42 and 30.43). However, these cases have to be closely monitored for any changes in the type of pain or

KEY BOX 30.19

PATHOGENESIS OF ADHESIONS



- Peritoneal plasminogen activator release results in lysis of adhesion.

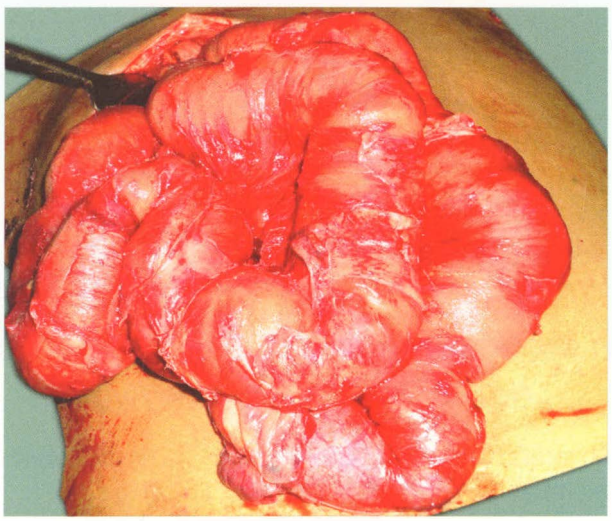


Fig. 30.37A: Bread and butter adhesions—case of perforation peritonitis

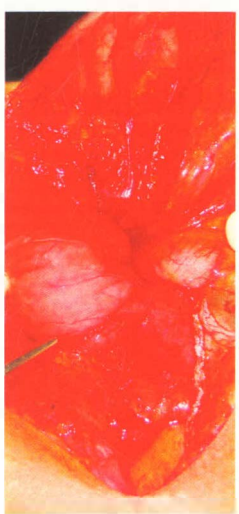


Fig. 30.37B: Band following tubectomy—has been divided

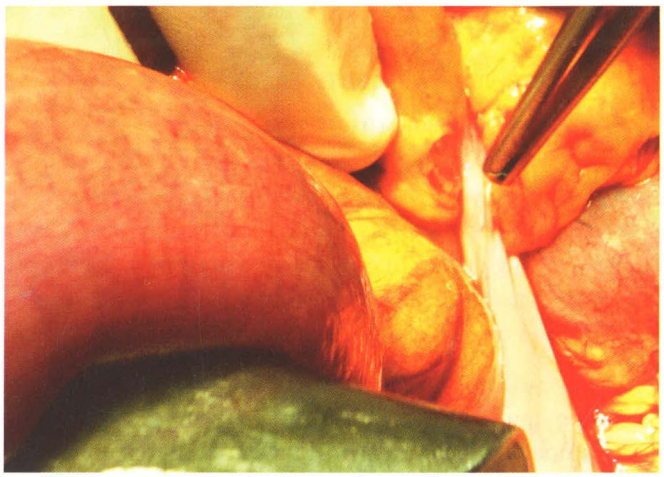


Fig. 30.38: Band causing obstruction — proximal dilated and distal collapsed bowel

Fig. 30.41: Congenital band causing obstruction

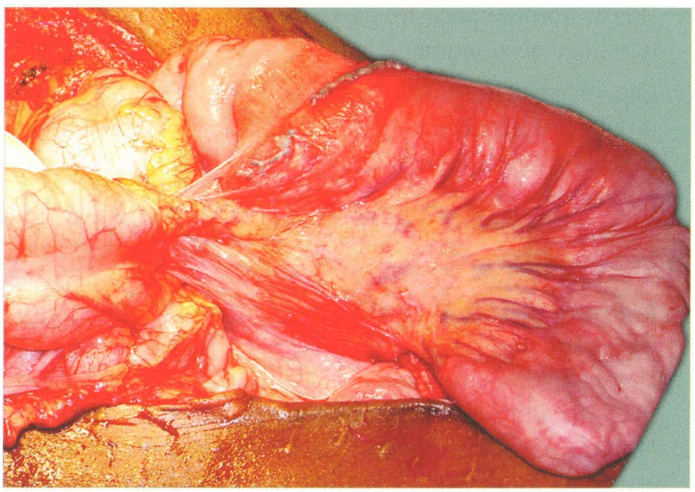


Fig. 30.39: Postappendicectomy band causing obstruction

Fig. 30.42: Adhesive obstruction

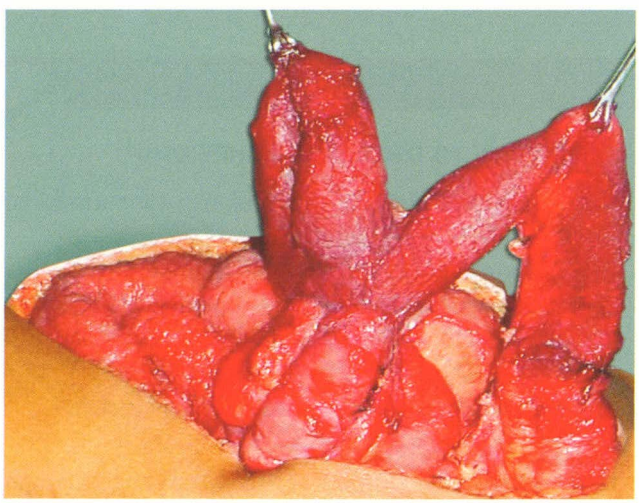


Fig. 30.40: Tuberculous adhesions and bands causing intestinal obstruction

Fig. 30.43: Close up view of gangrene

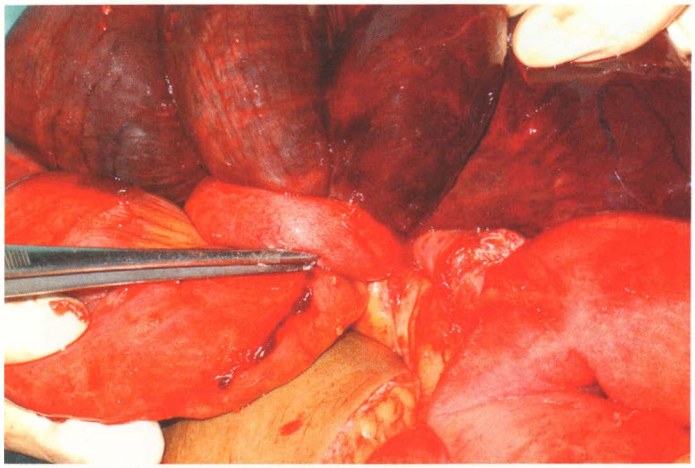


Fig. 30.44: Band secondary to peritonitis causing gangrene

KEY BOX 30.20

TYPES OF BAND

- Congenital : Transduodenal band of Ladd
  : Vitellointestinal duct associated band
- Acquired : Following peritonitis
  : Diverticulum
  : Greater omentum as a band

new abdominal signs. Severe pain, tachycardia, temperature, tachypnoea and tenderness in the abdomen indicates gangrene or perforation (Figs 30.43 and 30.44).

- **Gilroy Benan triad of adhesive pain:**
  - Pain gets aggravated or relieved on change of posture.
  - Pain in the region of old abdominal scar.
  - Tenderness elicited by pressure over the scar.

Investigations

- **Plain X-ray abdomen and small bowel enema** are very useful investigations to prove the obstruction.
- **Computed tomography (CT)** enhanced with oral contrast:
  - Detects air-fluid level: Complete obstruction
  - The absence of mass lesion
  - Dilated and collapsed loop junction
  - Thickening and oedema of the bowel wall suggests intestinal ischaemia. Presence of **intramural air** is a late sign (gangrene)
  - CT has a sensitivity of 90% and specificity of 88%
  - Thus, CT and MRI are very helpful in patients with small bowel obstruction.

Treatment

I. Conservative treatment

- In the form of nasogastric aspiration, IV replacement c fluids and electrolytes to correct dehydration may b successful in early postoperative obstruction. If it is nc successful, reoperation is required. Generally 48–72 hour is the waiting period in patients who present to the hospita as late adhesive obstruction. Further delay may result i perforation or gangrene of the bowel.
- Record pulse rate, blood pressure, abdominal girth an intake output. Increasing pulse rate, hypotension, increasin abdominal girth and oliguria in spite of adequate IV fluid will suggest gangrene. Such cases need to be explore immediately.

II. Surgical methods (Key Box 30.21)

- Where fibrous bands are the cause, they need to be divide to relieve obstruction.
- Laparoscopic adhesiolysis is more often being used and i is indicated in pelvic adhesion, selected cases of abdominal adhesion, single band adhesion and obstruction with mild distension.

III. Prevention of adhesion (Key Box 30.22)

A. Recently **absorbable and nonabsorbable membrane barriers** such as expanded polytetrafluoroethylene **(PTFE)** and membrane composed of hyaluronic acid and carboxymethyl cellulose have been used.

B. **Noble's plication:** By suturing loops together so that they are fixed in a suitable relation to one another (not very successful) (Fig. 30.45).

See Key Box 30.23 for summary of adhesive obstruction.

KEY BOX 30.21

HOW TO DECREASE ADHESIONS?

- Handle the bowel carefully. Good suturing without tension. Avoid anastomotic leak.
- Raw peritoneal areas should not be sutured
- Thorough peritoneal toilet in cases of peritonitis with saline or dextran to drain pus, bile, blood clots.
- Avoid spillage of contents—bile, faecal matter.
- Prefer a Pfannenstiel incision than midline incision.
- Noble's plication (Fig. 30.45)
- Laparoscopic method produces decreased adhesions than laparotomy.
- Membrane barriers

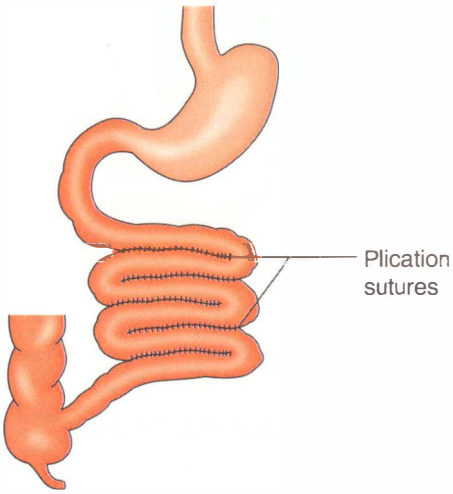


Fig. 30.45: Surgery for adhesions

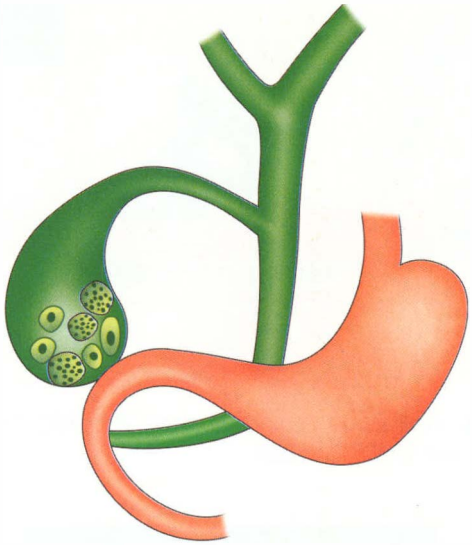


Fig. 30.46: Cholecystoduodenal fistula caused by gall stones

KEY BOX 30.22

MEMBRANE BARRIERS

- The bioresorbable membrane called seprafilm is currently the most effective membrane barrier.
- It consists of hyalorunic acid and carboxymethylcellulose
- At the completion of surgery, these films are placed at the potential sites of adhesion formation—such as pelvis, between the intestinal loops.
- Mechanism of action: Within the next 24 to 48 hours, the seprafilm membrane hydrates to form a gel-like barrier. It slowly resorbs within 7 days.
- These barriers should not be used to cover the anastomosis—chances of leak rates are high.

KEY BOX 30.23

ADHESIVE INTESTINAL OBSTRUCTION

- It is the most common cause of intestinal obstruction (in the West).
- The most common cause of adhesion is inflammatory—peritonitis.
- It is the cause of recurrent intestinal obstruction—often partial.
- Conservative treatment is successful in majority of cases.
- One can wait for 3–4 days with careful monitoring before a decision of laparotomy is undertaken (worsening situation).
- Repeated X-ray abdomen and CT scan will show changes and progression of the obstruction.
- Most valuable clinical signs of ischaemia/gangrene are tachycardia and tenderness.
- Adhesiolysis/resection is the treatment.

GALL STONE ILEUS—GALL STONE OBTURATION

Introduction: Gall stone ileus is more common in women and accounts for 1–4% of all presentations to hospital with small bowel obstruction. The term gall stone ileus is a misnomer, as the condition is a mechanical obstruction of the gut and not a true ileus.

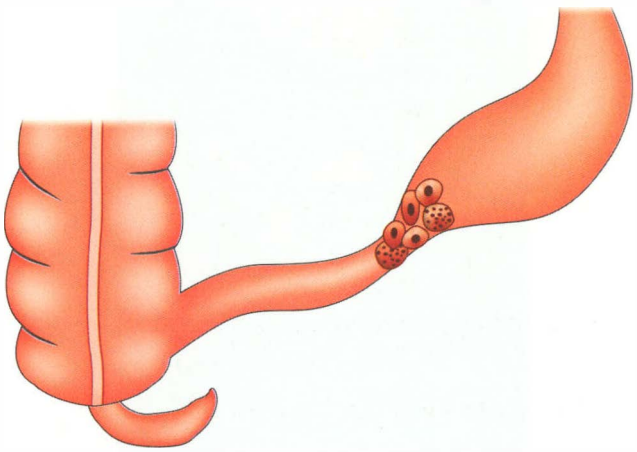


Fig. 30.47: Gall stones ileus (both these figures are contributed by Ms Vidhushi, Kasturba Medical College, Manipal)

- It should be suspected in a patient who has gall stones and present with intestinal obstruction.
- Elderly females above the age of 60 are usually affected.
- Gall stone reaches the terminal ileum by forming 'cholecystoduodenal fistula' due to recurrent attacks of cholecystitis (Figs 30.46 to 30.49).
- Due to recurrent inflammation, adhesions develop between gall bladder and duodenum (common) or gall bladder and colon or stomach (rare). Large stones cause pressure necrosis, resulting in formation of a cholecystoduodenal fistula.
- Terminal ileum is the narrowest portion of the gut wherein gall stone gets impacted. Sometimes, the stone may ulcerate from gall bladder into jejunum, colon, etc.

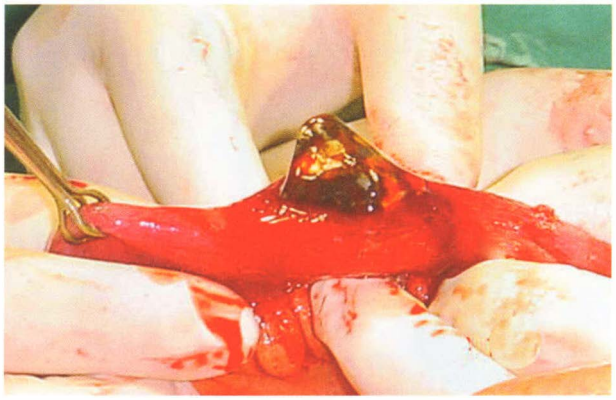


Fig. 30.48: Gall stone ileus (*Courtesy:* Prof MG Shenoy and Dr GN Prasad, KMC, Manipal)

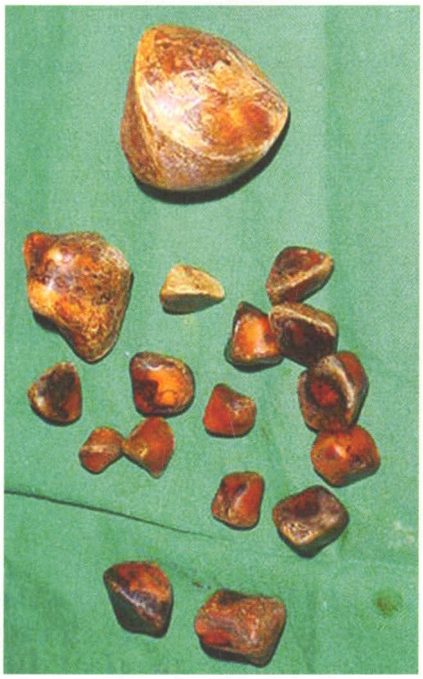


Fig. 30.49: 17 Gall stones were removed in this case

- ***Clinical features:*** They are suggestive of small intestinal obstruction—abdominal pain is severe, vomiting and distension. Step ladder peristalsis may be seen. History suggestive of recurrent cholecystitis may be present.
- ***Rigler's triad:*** Pneumobilia, the presence of an aberrant gall stone and enteric obstruction (Fig. 30.50).

  Bouveret syndrome: It is a gastric outlet syndrome secondary to stone lodged in proximal duodenum due to cholecystoduodenal fistula. Proximal migration of the stone will precipitate obstruction.

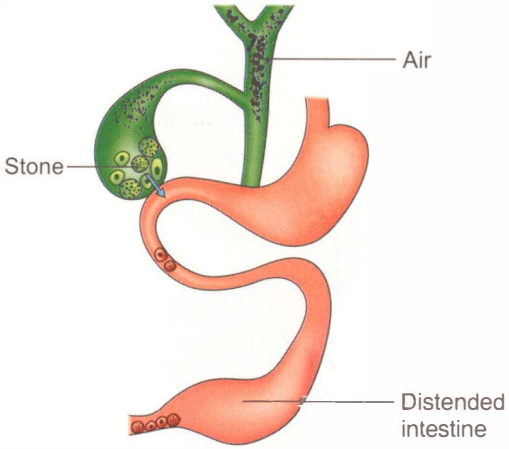


Fig. 30.50: Rigler's triad

Investigations

1. **Plain X-ray abdomen** (erect position) may demonstrate multiple gas and fluid levels and stone in the gall bladder and also in the lower abdomen suggesting gall stone ileus.
   - **Air may be found**[1] in the biliary system.
2. **Small bowel enema** may demonstrate partial obstruction.
3. **Computed tomography (CT)** scanning is the investigation of choice. It has preoperative diagnosis of gall stone ileus with a sensitivity of 93%. It will also indicate any inflammation of the gall bladder, fistula-air pockets in the gall bladder, distal stone and proximal dilatation.

Treatment

- There is difference of opinion regarding what is the ideal surgery to be done in case of gall stone obstruction. It is

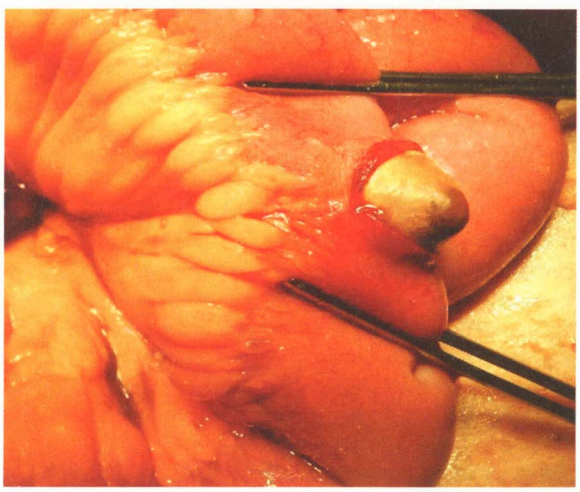


Fig. 30.51: Enterolithotomy and removal of stone

[1]Cholecystoduodenal fistula, choledochoduodenostomy, sphincteroplasty, emphysematous cholecystitis are a few other conditions wherein air is found within biliary tree.

because many patients are elderly, with co-morbidity and they have intestinal obstruction (dehydration, bowel oedema, bacterial proliferation, etc). Whatever it is, the first step is always to do enterolithotomy by incising the ileum and deliver the stone/stones. It may be possible to crush the stone and pass it onto caecum, avoiding enterolithotomy. Search the proximal intestine for any other stones. What must be done next? Two types are described.

1. **Single stage procedure:** If general condition of the patient is good, and gall bladder is gangrenous or inflamed (more chances of perforation) enterolithotomy, cholecystectomy followed by closure of the duodenal fistula is done.
2. **Two stage procedure:** If general condition is not good or with no active inflammation of the gall bladder, enterolithotomy is the procedure of choice, followed 6 weeks later by cholecystectomy with closure of the duodenal fistula (Fig. 30.51).

INTUSSUSCEPTION

Definition

Invagination of one segment of intestine into another (usually the proximal into distal) is called intussusception.

PEARLS OF WISDOM

It is the most common cause of intestinal obstruction in infants aged 6 to 18 months.

Incidence: 2–4/1000 live births.

Types

1. **Simple ileocolic** is the most common type, followed by ileoileal or colocolic.
2. **Compound**—ileoileocolic
3. **Retrograde jejunogastric intussusception**, a complication of gastrojejunostomy (GJ) is a rare but interesting type of intussusception (*see* page 503).

Parts (Fig. 30.52)

1. **Intussuscipiens:** It is the outer tube (distal bowel which receives the intestine).
2. **Intussusceptum:** Proximal bowel (inner tube) which enters the distal segment.
3. **Apex** is the part which advances further into the distal bowel.
4. **Neck**, the narrowest portion of intussusception, is the junction of entering layer with the mass.
   - The whole mass that develops is called intussusception.

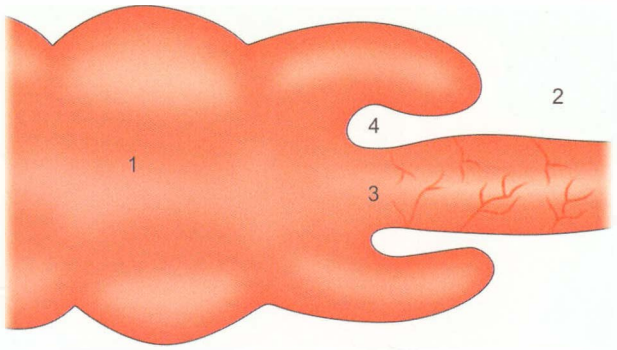


Fig. 30.52: Parts of intussusception (*see* the text for numbers)

AETIOPATHOGENESIS

1. **Idiopathic intussusception:** Actual cause is not known. It is seen in infants. Possible factors (Key Box 30.24).
   - **Dietary factor:** Around the age of 6–9 months, weaning of breast milk is done. Weaning causes alteration in the bacterial flora in the GIT, causing enlargement of the Peyer's patches. These protrude into the terminal ileum and may precipitate intussusception.

KEY BOX 30.24

PREDISPOSING FACTORS

- Recent viral infection (upper respiratory)
- Recent operation
- Henöch-Schönlein purpura
- Cystic fibrosis
- Coeliac disease
- Haemophilia

   - **Infective factor:** It usually follows upper respiratory tract infection with virus (adenorotaviruses) which produce inflammation of Peyer's patches.
2. **Adult intussusception (secondary):** In adults, there is always a cause for intussusception (Key Box 30.25 and Figs 30.53 to 30.62).

KEY BOX 30.25

ADULT INTUSSUSCEPTION

- Meckel's diverticulum
- Polyps, tumours, submucous lipoma
- Carcinoma colon (caecum, transverse colon)

Pathophysiology

- As the apex advances, it drags the mesentery containing blood vessels which get obstructed at the neck resulting in mucosal ulcers and haemorrhages. Marked lymphadenopathy and hypertrophy of Peyer's patch is found at operation.
- If the neck is too tight, gangrene sets in very early, as in ileocolic intussusception.
- All other features of strangulation, dehydration, distension and septicaemic shock develop later.

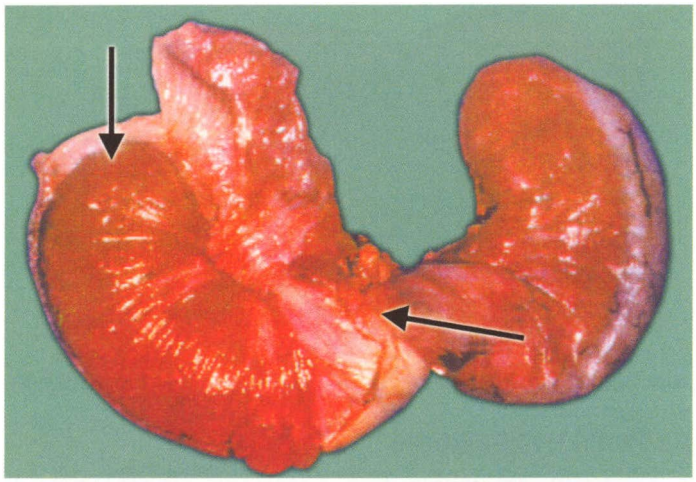


Fig. 30.53: Intussusception

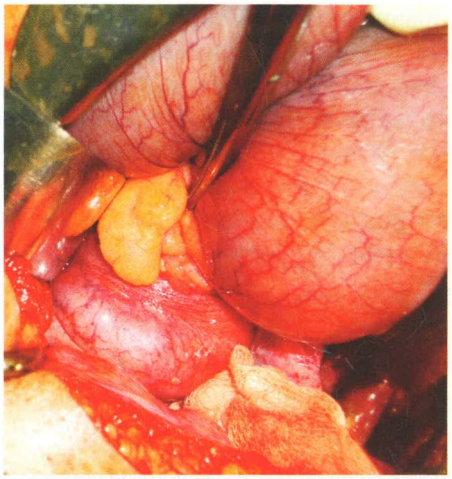


Fig. 30.54: Carcinoid of the ileum with intussusception

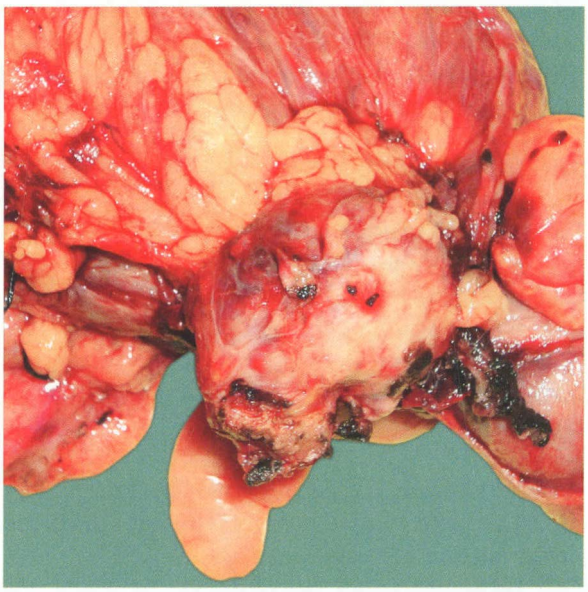


Fig. 30.55: Carcinoid of the ileum causing intussusception—resected specimen

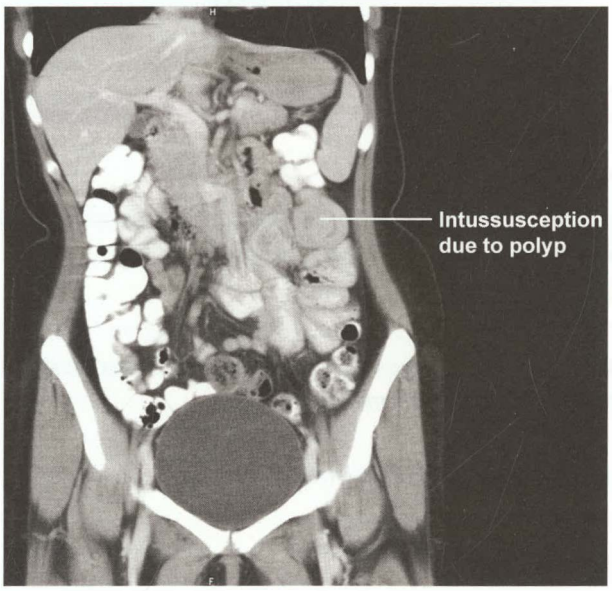


Fig. 30.56: Intussusception due to polyp

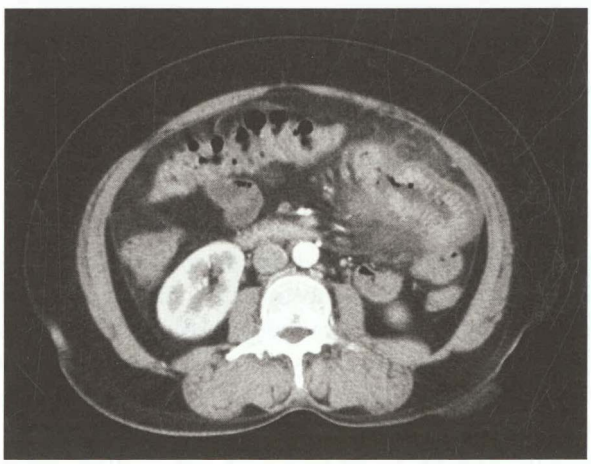


Fig. 30.57: Jejunal carcinoma—CT coronal section

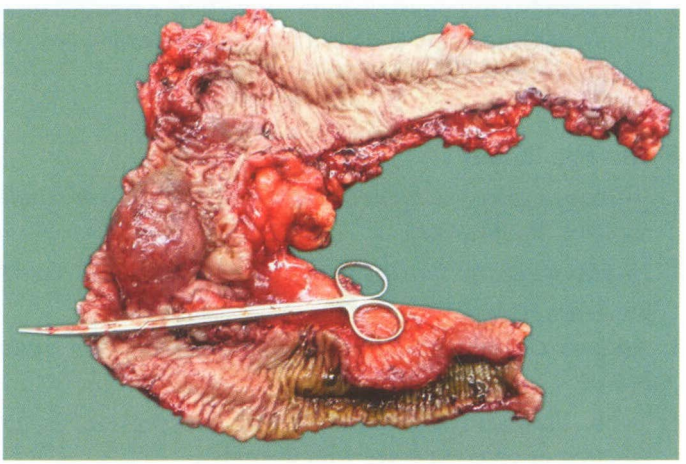


Fig. 30.58: Opened specimen showing polyp

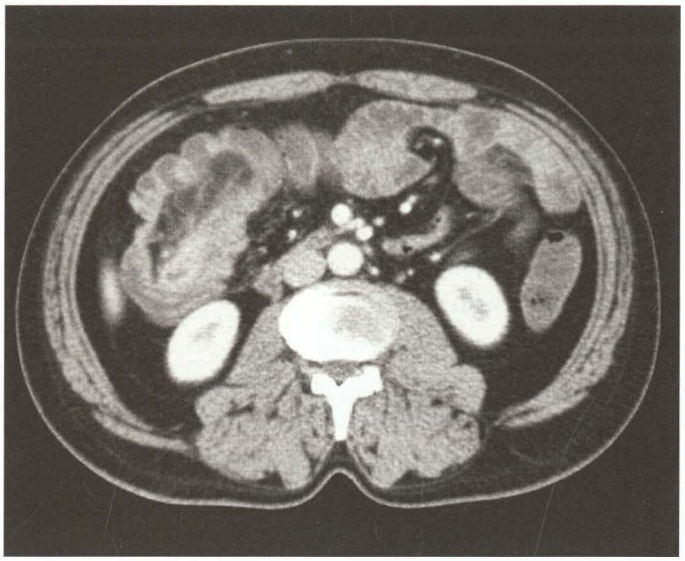


Fig. 30.59: Ileocolic intussusception CT

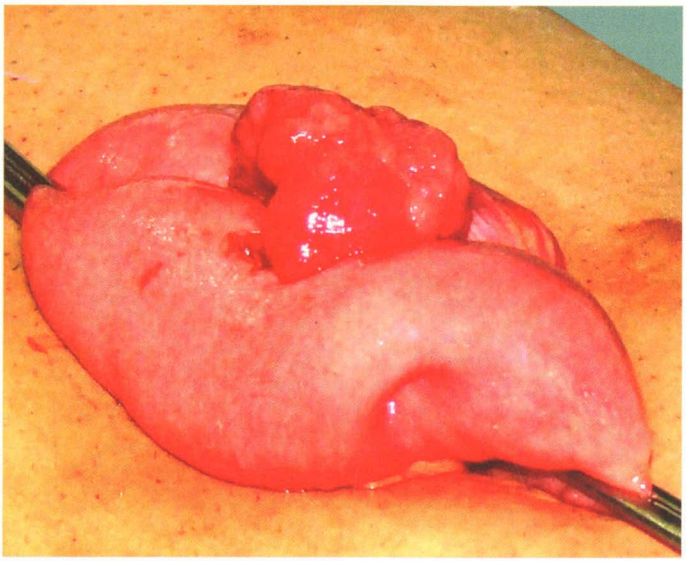


Fig. 30.60: Intussusception due to polyp

Fig. 30.61: Intussusception due to polyp at surgery

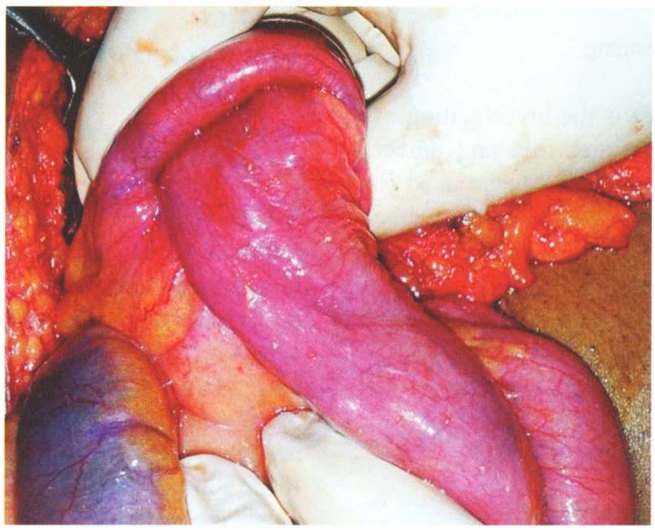


Fig. 30.62: Ileocolic intussusception at surgery

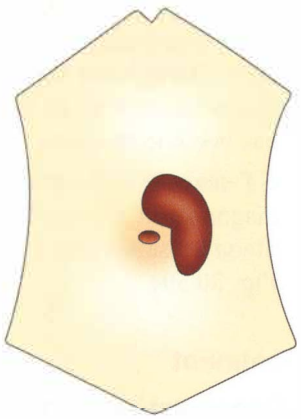


Fig. 30.63: Sausage-shaped mass—resonant, mobile

Clinical features

- First born male infants between 6 and 9 months are commonly affected. Boys : Girls — 3 : 2.
- **Child screams** with abdominal pain[1] (intestinal colic) which is associated with facial pallor.
- One attack of **red currant jelly stools** is characteristic. Bleeding is due to mucosal ulcer (venous infarction). Mucus secretion is due to irritation of intestines. This is followed by absolute constipation. **Red currant jelly stools are not found in adult intussusception.**
- Vomiting 3–4 times, initially due to pylorospasm. Later, due to obstruction.
- In between the spasms, the child sleeps but gets up suddenly with pain.

Signs

- The mother is asked to feed the baby in sitting position and examination of the baby's abdomen is done with the left hand, standing in front of the mother.
- A contracting, hardening mass in and around the umbilical region can be felt **(sausage-shaped,** Fig. 30.63).
- **Emptiness in the right iliac fossa** (Dance's sign—*signe de dance*).
- There may be a **visible step ladder peristalsis**.
- Rarely, intussusception can be seen **outside the anus** due to long mesentery.
- Rectal examination reveals **blood-stained mucus** on the examining finger.
- Features of peritonitis occur in untreated cases.

[1]Hindi speaking mother says, '*Baccha sota hai aur rota hai*'

Investigations

- **Barium enema:** 'Claw (pincer) ending' (Fig. 30.64) is diagnostic of intussusception. This is also called '**meniscus sign**'. If there is any suspicion of gangrene, this test should not be done. In many cases, the diagnosis is established on clinical grounds.

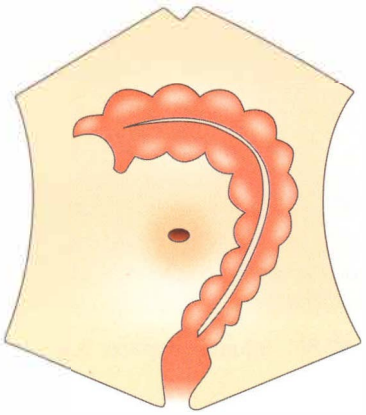


Fig. 30.64: Claw ending

- Ultrasound (Figs 30.65 and 30.66) is the investigation of choice nowadays. It can detect target sign and detect mass **(Doughnut sign)** with Doppler. It can be used to assess vascularity of the bowel also. Thus, barium enema has become obsolete.
- **CT scan:** It is the most sensitive imaging modality in the diagnosis of intussusception. Sausage-shaped mass, with blood vessels within bowel lumen are typical findings (*see* Fig. 30.59).

Treatment

I. Conservative treatment (Key Box 30.26)

- Hydrostatic reduction can be attempted when the gangrene is ruled out as in early intussusception. A lubricated catheter is introduced into the rectum and 1–2 litres of saline from a height of 1–2 metres is allowed to run. Catheter is removed and buttocks are pressed together. 50–70% of cases are reduced by this method, 1:3 barium sulphate in warm isotonic saline can also be used.
- Air contrast enema will not reduce gangrenous bowel. Air is pumped into the colon at a pressure of 60–80 mmHg.

Contraindications:

- Peritonitis with shock
- Total intestinal obstruction

KEY BOX 30.26

HYDROSTATIC REDUCTION IS SUCCESSFUL WHEN

1. Flatus and faeces are passed with barium
2. Child is symptom-free and comfortable
3. Small bowel loops are filled with contrast

Advantage

- Easy, nonoperative method

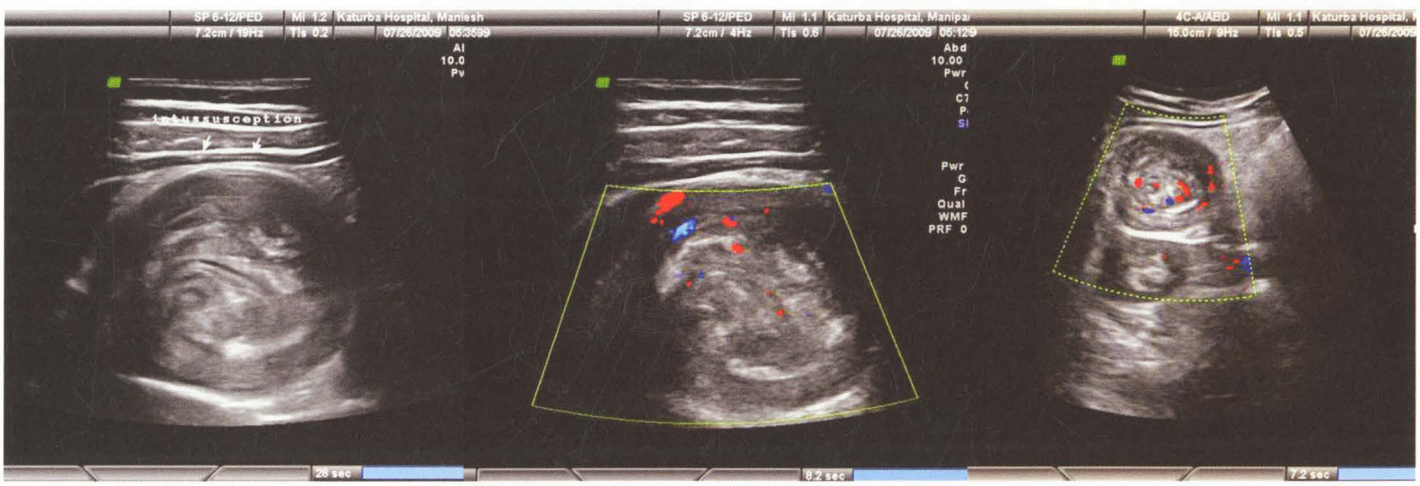
Complication

- Rarely, colonic perforation

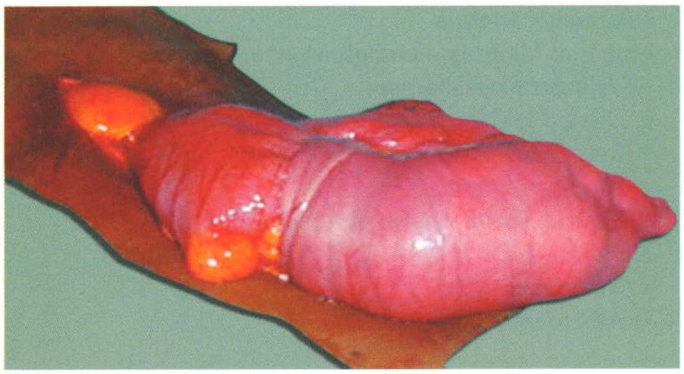
II. Surgical treatment (Figs 30.67 to 30.69)

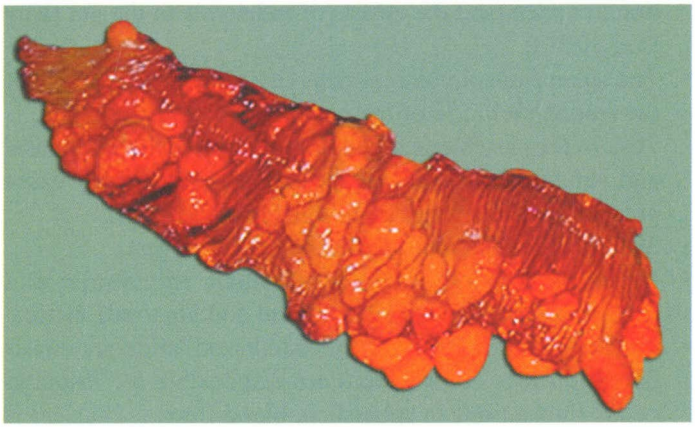
Laparotomy and reduction of intussusception

- Intussusception is reduced by milking (squeezing) the colon in opposite direction, which is facilitated by breaking the adhesions at the neck using the little finger. Appendicectomy is also done, as it avoids any future confusion as to the reason for the abdominal scar. Fixing the caecum is not necessary because idiopathic intussusception rarely recurs. If the loop is gangrenous, resection and ileocolic anastomosis is done.
- **Recurrent intussusception is rare:** If it occurs, terminal ileum is sutured to the side of the ascending colon.



Figs 30.65 and 30.66: Ultrasound—pseudokidney sign, target sign, Duplex—assesses vascularity also





Figs 30.67 and 30.68: Adult intussusception due to jejunal lipomatosis. This patient was 24-year-old male who presented with intestinal obstruction. A mass was palpable in the umbilical region at laparotomy. This mass was resected. Opened specimen showed extensive segmental lipomatosis (*Courtesy:* Dr Gabriel Rodrigues, Dr Mahesh Gopa Setty, Dr Lavanya K, KMC, Manipal)

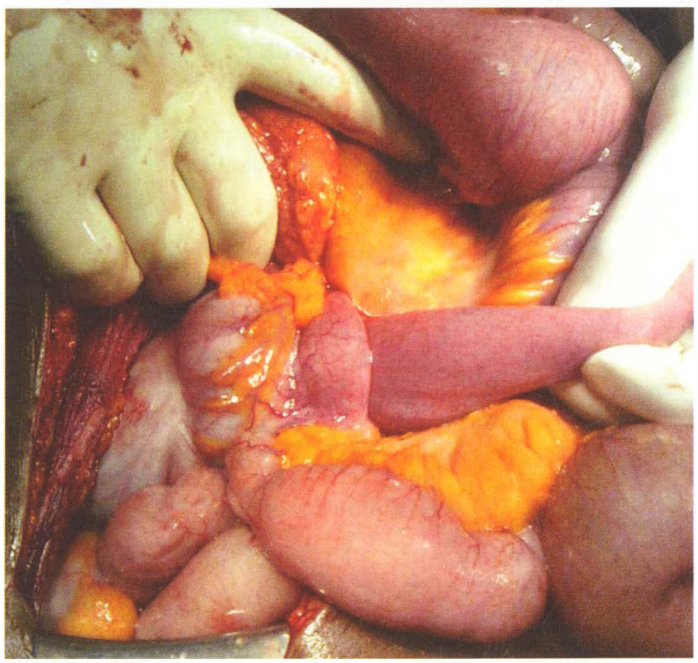


Fig. 30.69: Intussusception is reduced at surgery

See Key Box 30.27 for some interesting "more common" about intussusception.

KEY BOX 30.27

ACUTE INTUSSUSCEPTION—MOST COMMON

- Most common in children between 5 and 10 months of age
- Most common cause of intestinal obstruction in children
- Most common cause is idiopathic (90%)
- Most incidence of adenovirus infection, common in midsummer and midwinter
- Most common variety is ileocolic variety
- Most commonly hypertrophied Peyer's patches
- Most common type of nonoperative reduction is by using air and barium enema
- Most commonly done surgical procedure is reduction
- Most commonly used noninvasive test for diagnosis is ultrasound—pseudokidney sign, target sign, Duplex-assesses vascularity also.

MESENTERIC VASCULAR OCCLUSION

Definition

Acute mesenteric ischaemia is an abrupt reduction in blood flow to the intestinal circulation of sufficient magnitude to compromise the metabolic requirements and potentially threaten the viability of the affected organs (Key Box 30.28).

Clinical features

- It is common in elderly patients who are hypertensive and usually obese.
- It is due to atherosclerosis causing thrombosis of the superior mesenteric artery or due to emboli which originate from atheromatous plaques or from the infarcted heart.
- Smokers are more often affected by this condition.
- **Superior mesenteric vein** can also get thrombosed due to injury during pancreatectomy or thrombosis as a result of oral contraceptive pills.

KEY BOX 30.28

TYPES

1. Acute mesenteric ischaemia (AMI): It is a sudden occlusion of artery or vein resulting in gangrene. It has a mortality rate of 60 to 90%.
2. Chronic mesenteric ischaemia (CMI): It is associated with stenosis of coeliac artery, superior mesenteric artery or inferior mesenteric artery.

Effects

The pathological effects of arterial occlusion and venous occlusion are the same. Superior mesenteric artery supplies the entire midgut starting from the duodenojejunal flexure to right one-third of the transverse colon. It is an end-artery. As a result of thrombosis, the entire small bowel and portion of the large bowel becomes gangrenous (if there is a thrombus at the origin of superior mesenteric artery).

Causes of acute mesenteric ischaemia

1. **Arterial emboli** are the most frequent cause of AMI and are responsible for approximately 40 to 50% of cases. Most mesenteric emboli originate from a cardiac source. They lodge in the superior mesenteric artery (SMA) because it emerges from the aorta at an oblique angle. Few emboli lodge in the origin of middle colic artery.
2. **Acute mesenteric thrombosis** accounts for 25 to 30% of all ischaemic cases. Atherosclerotic disease is the cause of thrombosis. It typically occurs at the origin of the superior mesenteric artery. Gangrene is more extensive in cases of thrombosis than embolism.
3. **Nonocclusive mesenteric ischaemia (NOMI):** Typically happens in cases of hypotensive patients. Low cardiac output, ICU patients on vasoconstrictors and inotropes precipitate the problem. Splanchnic vasoconstriction occur in response to hypovolemia. Vasoactive drugs, particularly digoxin, have been implicated in the pathogenesis of NOMI.
4. **Mesenteric venous thrombosis (MVT)** is the least common cause of mesenteric ischaemia, representing up to 10% of all patients with mesenteric ischaemia related to primary clotting disorders. Thrombi usually originate in the venous arcades and propagate (Key Box 30.29).

KEY BOX 30.29

AETIOLOGY OF MESENTERIC VENOUS THROMBOSIS

- Hypercoagulable states (e.g. polycythaemia vera, protein C and S deficiencies).
- Visceral infection
- Portal hypertension
- Blunt abdominal trauma
- Malignancy: Pancreatic
- Pancreatitis and in patients who smoke
- Women taking oral contraceptives are also at increased risk of venous thrombosis.
- Patients who have undergone splenectomy.

To summarise the important causes

1. Atherosclerosis: Hypotension, haemorrhage into atheroma
2. Hypercoagulable state
3. Fibromuscular dysplasia
4. Takayasu's arteritis
5. Embolic: Majority—emboli originate in the heart
   - Valvular heart disease
   - Dilated LA
   - Recent MI
   - Atrial arrhythmias
   - Ventricular dilatation with mural thrombus
6. Aorta:
   - Aneurysm
   - Atheroemboli

Splanchnic circulation

- It is better to have some idea about the splanchnic circulation to understand the effects of ischaemia to the gut (Key Box 30.30).
- The splanchnic circulation receives approximately 25% of the resting and 35% of the postprandial cardiac output.
- 70% of the mesenteric blood flow is directed to the mucosal and submucosal layers of the bowel. Hence early cases present with bleeding due to mucosal ulcerations.
- 30% supplies the muscularis and serosal layers.
- Blood flow is regulated by intrinsic (metabolic and myogenic) and the extrinsic (neural and humoral) factors.
- Reactive hyperaemia, and hypoxic vasodilation are considered intrinsic controls and are responsible for instantaneous fluctuations in splanchnic blood flow.
- An imbalance between tissue oxygen supply and demand will raise the concentration of local metabolites (e.g. hydrogen, potassium, carbon dioxide, and adenosine), resulting in vasodilation and hyperaemia.

KEY BOX 30.30

ISCHAEMIC INJURY

Vascular occlusion

↓

Inflammatory cell infiltrate

↓

Loss of capillary integrity

↓

Bowel wall oedema

↓

Breakdown of mucosal barrier

↓

Bacterial translocation

↓

Endotoxaemia

↓

Sequestration of fluid within bowel lumen

Reperfusion injury

- Oxygen free radicals
- Cytokines from PMN leucocytes
- Myocardial depression
- Progressive inflammatory response
- Increased capillary permeability
- Tissue oedema and MODS.

Clinical features

Symptoms

1. **Abdominal pain:** Severe, poorly localised, unresponsive to narcotics, out of proportion to the physical findings (Fig. 30.70).
2. Onset may be abrupt (embolism) or insidious (thrombosis)
3. **Gastrointestinal emptying:** Vomiting, diarrhoea, with occult or frank bleeding once infarction set in.

Signs

1. **Early:** Soft, nontender, normal bowel sounds.
2. **Late:** Distension, tenderness, rebound tenderness, rigidity, absent bowel sounds.
3. **Terminal:** Cold clammy extremities––dehydration, thready pulse, hypotension, shock, acidosis.

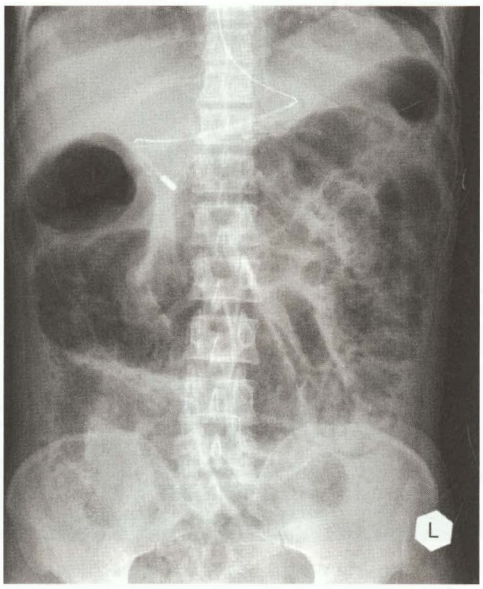
PEARLS OF WISDOM

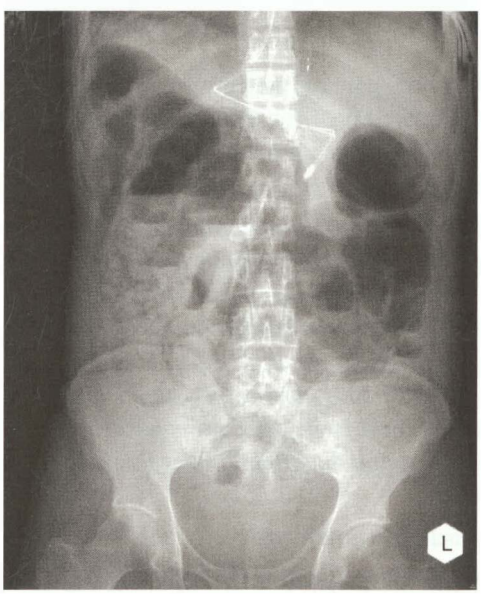
Sudden event, sudden bleeding, sudden collapse, severe pain, silent abdomen, shock progressing to sepsis is mesenteric ischaemia.

Investigations

1. **Total counts** are raised, in about 75% of cases > 15,000 cells/mm^3.
2. **Plain X-ray abdomen** (erect) reveals absence of gas within the bowel loops and intramural gas. Blunt plicae are seen. This is called as thumb printing (Figs 30.71 and 30.72).
3. **Metabolic acidosis** in > 50% of cases.
4. **Serum phosphate** levels are raised within 3–4 hours following ischaemia as smooth muscle layer of small bowel is rich in phosphates.
5. **D-dimer** is elevated in all cases.
6. **CT with or without angiogram** is the investigation of choice. It can detect **intramural air pockets** in the bowel wall, air within biliary radicle, perforation, etc. It can also detect thrombus or narrowing of superior mesenteric artery or superior mesenteric vein (Figs 30.73 and 30.74).
7. **Emergency angiography** is the test of choice and can be done within 6 hours of ischaemia. It can demonstrate arterial thrombosis at the origin or distally in one of the branches. (It can also be therapeutic.)
8. **Hypercoagulability disorders** — protein C and protein S antithrombin III, factor V, anticardiolipin antibody (MVT).

Fig. 30.70: Attitude of a patient with severe ischaemia due to superior mesenteric vascular occlusion. In these cases, the pain is disproportionate to the abdominal signs





Figs 30.71 and 30.72: Plain X-ray showing presence of air in the bowel wall

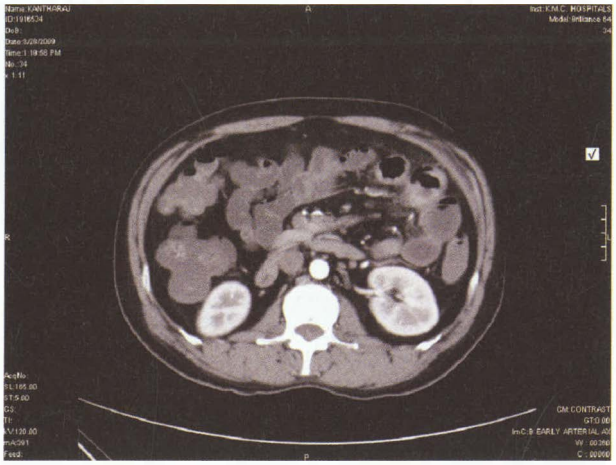


Fig. 30.73: CT showing SMA thrombosis

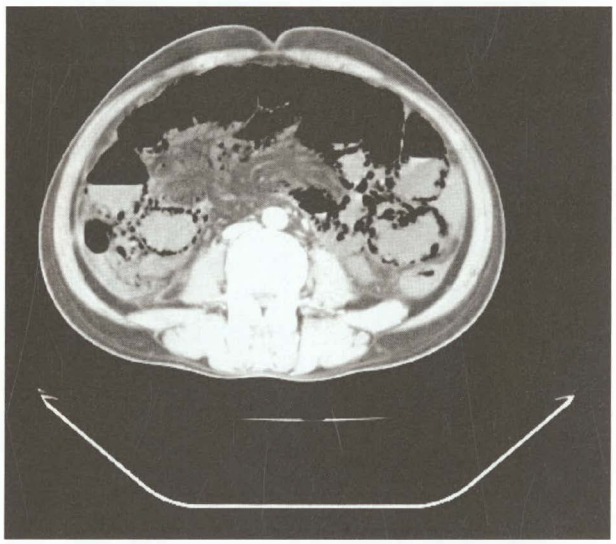


Fig. 30.74: CT confirming the same. A case of superior mesenteric arterial thrombosis with massive gangrene (*See* Fig. 30.75)

Treatment

1. Majority of the patients present late with massive gangrene. Massive resection of the gangrenous bowel followed by end to end anastomosis is done. These patients suffer from short bowel syndrome *(vide infra)*, if they survive.
2. If patients come within 4–6 hours of ischaemia, emergency angiography followed by papaverine infusion (30 to 60 mg/h) into the superior mesenteric artery can be tried. Otherwise, emergency laparotomy is done and the superior mesenteric artery is explored. A Fogarty catheter is introduced and embolectomy is done. These patients may require a **second look operation within 24–48 hours** to rule out gangrene developing later due to rethrombosis of the artery.
3. Heparinisation—in all cases
4. Other vasodilator agents used are:
   a. Tolazoline
   b. Glucagon
   c. Nitroglycerine
   d. Nitroprusside
   e. Prostaglandin E
   f. Phenoxybenzamine
   g. Isoproterenol
5. Sympathetic epidural block

Assessing bowel viability

- Doppler (84%)
  - Absent arterial flow on antimesenteric border of bowel.
  - Absent mesenteric arterial flow
- Fluorescein test (100%)
  - Sodium fluorescein 1 g administered IV over 30 to 60 seconds and the bowel examined using a hand-held long wave UV Wood's lamp (yellow).

Recent advances in cases of mesenteric ischaemia

- Catheter directed thrombolysis
- Percutaneous transluminal angioplasty
- Endovascular fenestration of aortic dissection:
  - Techniques to assess *bowel viability*
  - Pulse oximetry
  - Infrared photoplethysmography
  - Bowel surface oximetry
  - Quantitative fluorescence using perfusion fluorometer.

Prognosis

Majority of the cases present with massive gangrene. Even after massive resection, they succumb to the sepsis and multiorgan failure (Fig. 30.75).

See next page for a few causes of intestinal obstruction (Figs 30.76 to 30.81).

Fig. 30.75: Massive gangrene of small intestines and right side of the colon due to thrombosis at the origin of superior mesenteric artery

A FEW CAUSES OF INTESTINAL OBSTRUCTION

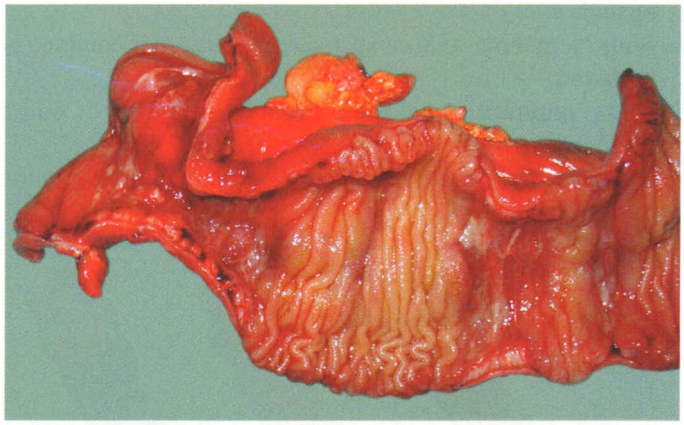


Fig. 30.76: Tubercular stricture—annular stricture

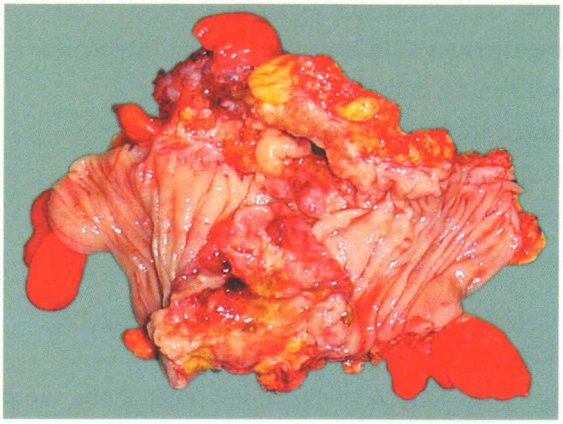
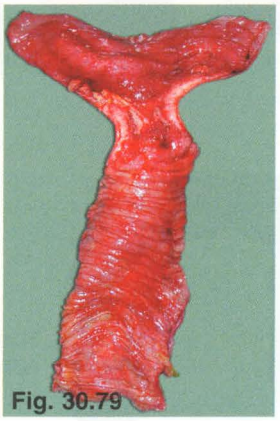
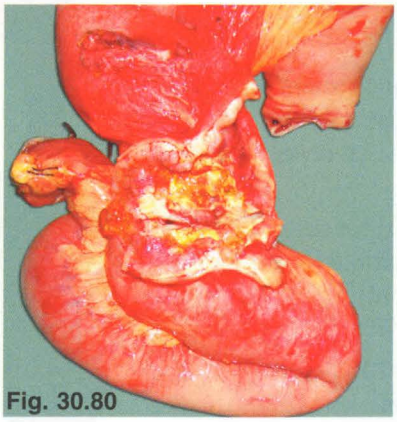


Fig. 30.77: Sigmoid stricture due to carcinoma





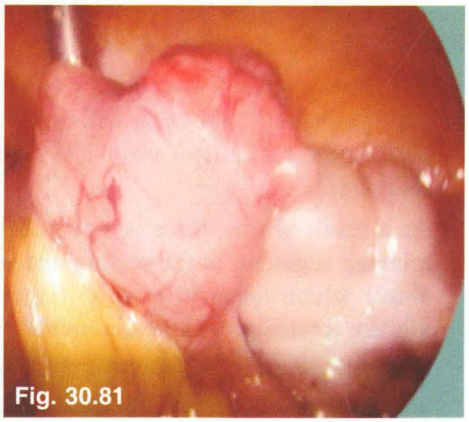


Fig. 30.78: Obstructed hernia **Fig. 30.79:** Nonspecific stricture **Fig. 30.80:** Peritoneal metastasis causing obstruction opened lesion
Fig. 30.81: Laparoscopy detected a lesion in the small intestine

STRICTURES

- Common causes are tubercular stricture of the ileum or jejunum in India and Crohn's disease in the Western world.
- Radiation stricture, ischaemic strictures and nonspecific strictures are the other causes.
- Small bowel enema or enteroscopy are very useful investigations (Figs 30.82 to 30.85).
- Malignant structures

Fig. 30.82: Enteroclysis showing jejunal narrowing and aortic calcification

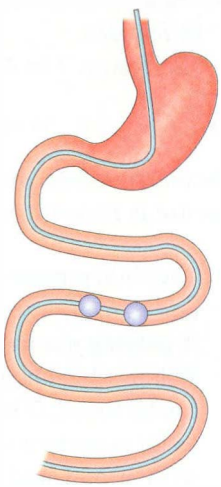


Fig. 30.83: Double balloon enteroscopy representation

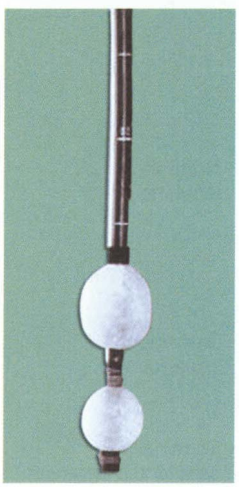


Fig. 30.84: Double balloon enteroscopy

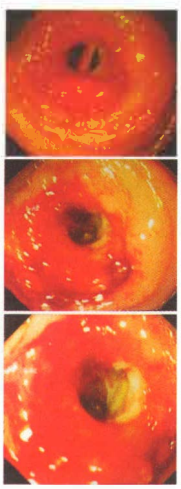


Fig. 30.85: Balloon enteroscopy showing jejunal ulcers

NEONATAL INTESTINAL OBSTRUCTION

Causes

1. **Hirschsprung's disease**—congenital megacolon
2. Atresia and stenosis
3. Arrested rotation with bands
4. Volvulus neonatorum
5. Meconium ileus
6. Imperforate anus

HIRSCHSPRUNG'S DISEASE: CONGENITAL MEGACOLON

It is also called congenital megacolon, aganglionic megacolon or primary megacolon. It is one of the common causes of neonatal intestinal obstruction.

Pathophysiology (Fig. 30.86)

- The disease always involves the anus and rectum wherein parasympathetic ganglion cells are absent in the neural plexus of the intestinal wall. The defect involves internal sphincter.
- As a result of this, there is a terminally constricted, non-relaxing segment, in the rectum and sigmoid (lower part), above which the pelvic colon (sigmoid) is enormously dilated. Rectosigmoid area is involved in 80% of cases.
- Circular muscle hypertrophy, mucosal hyperaemia and ulcers are present in the dilated segments.
- In between, there may be a transition zone (cone), which contains a few parasympathetic ganglion cells.
- Rarely, Hirschsprung's can also involve the entire sigmoid colon or even the entire colon.
- Hirschsprung's rarely occurs in adults also.

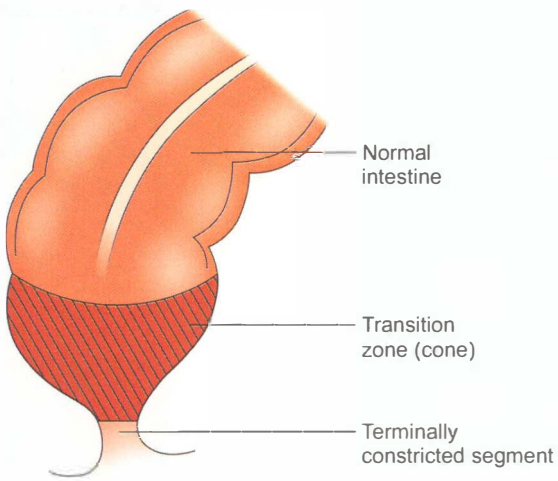


Fig. 30.86: Pathophysiology

Types of Hirschsprung's disease (Fig. 30.87)

1. **Ultrashort segment:** Anal canal and terminal rectum is aganglionic.
2. **Short segment:** Anal canal and entire rectum is completely involved.
3. **Long segment:** Anal canal, rectum and part of colon involved.
4. **Total colonic:** Anal canal, rectum and whole length of colon is involved.

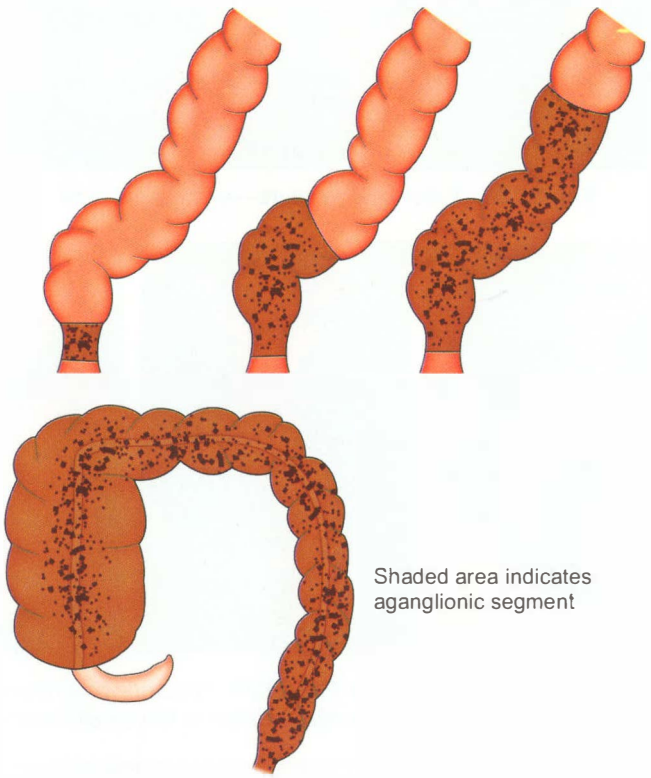


Fig. 30.87: Types of Hirschsprung's disease

Clinical features

- Male children are commonly affected, when compared to females.
- **Incidence:** 1 in 4000 to 5000 live births.

PEARLS OF WISDOM

The most common associated anomaly with Hirschsprung's disease is Down's syndrome (5 to 10%).

- The child presents with acute neonatal intestinal obstruction as manifested by failure to pass meconium or delay in passing meconium with abdominal distension.
- Within 12–24 hours, all features of intestinal obstruction can be found. If it is complicated by enterocolitis, it may result in perforation and septicaemia. A severe diarrhoea with blood and mucus, abdominal distension and vomiting can occur within a few hours, followed by hypovolaemic shock.

- Rectal examination reveals that the rectum is empty, finger is gripped by anal sphincter and there is no perianal soiling (Fig. 30.88). On the other hand, in acquired megacolon, rectum is loaded with faecal matter, perianal soiling is present and there is no sphincter activity (Fig. 30.89).
- **Chronic variety:** Chronic constipation manifesting in the first few weeks of life. The child may be brought with abdominal distension. Stools are goat pellet-like.

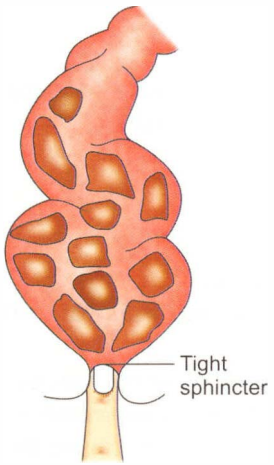


Fig. 30.88: Rectal examination in congenital megacolon

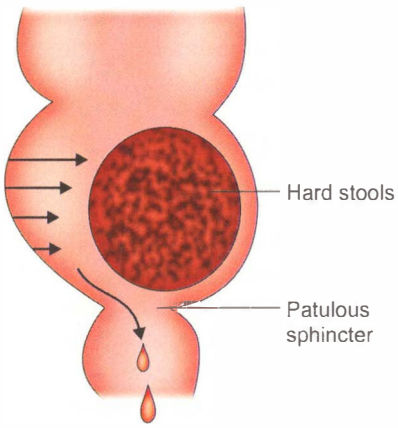


Fig. 30.89: Acquired megacolon

Differential diagnosis (Key Box 30.31)

- **Acquired megacolon:** Usually manifests by one to two years of age. Rectum is loaded with faecal matter.

Complications

- Intestinal obstruction, perforation, peritonitis
- Enterocolitis
- Growth retardation

Investigations

1. **Full thickness rectal wall** biopsy under GA demonstrates absence of parasympathetic ganglion cells and hypertrophic nerve fibres in the nerve plexus. It should be taken above the anorectal junction.

PEARLS OF WISDOM

Today submucosal suction biopsy is more popular than biopsy since it avoids haemorrhage, infection and scarring.

2. **Barium enema:** 3.6% solution of barium is used, the intermediate zone appears as a cone with proximal dilatation and a distal narrow zone which is characteristic of Hirschsprung's disease.

Treatment

I. Emergency cases

- Right transverse loop colostomy: In most of the cases, aganglionic segment is limited to rectosigmoid region.

KEY BOX 30.31

DIFFERENTIAL DIAGNOSIS

- Hypothyroidism
- Meconium plug syndrome
- Intestinal pseudo-obstruction
- Colonic neuronal dysplasia

- A full thickness biopsy of the colostomy is sent for histopathological examination.

II. Definitive surgery

- Can be done usually between the age of 3 and 6 months (8 to 10 kg of weight).
- Resection of aganglionic bowel (anorectum) followed by a pull-through procedure. **Maintaining continence is the main aim.**
- A few points of comparison between 'Duhamel's and Swenson's pull-through' are mentioned below.

Duhamel's	Swenson's
1. Retrorectal pull-through	I. Endorectal pull-through
2. Technically easy	2. Difficult

Steps of Duhamel's pull-through surgery

1. The rectum is transected above the peritoneal fold and is closed.
2. The proximal ganglionic segment is pulled down behind the rectum (retrorectal space created by using blunt dissector).
3. An incision is made in the posterior wall of the anorectum above the dentate line and is deepened through the entire bowel wall.
4. The end of the proximal colon is sutured to the opening in the posterior anal canal all around.
5. The adjacent walls of rectum (posterior wall) and colon (anterior wall) are crushed by using a Kocher's forceps which falls off by itself by the 14th day.
6. The open end of the rectum (above) is closed end to side to the colon.

Other types of surgery: Soave's mucosectomy and pull-through operation.

ATRESIA AND STENOSIS

- Commonly, it affects the duodenum, followed by ileum and jejunum (Fig. 30.90).
- There may be single/multiple atresia.
- Incidence: 1 in 10,000 live births.

PEARLS OF WISDOM

Duodenal atresia is the most common cause of intestinal obstruction in neonates.

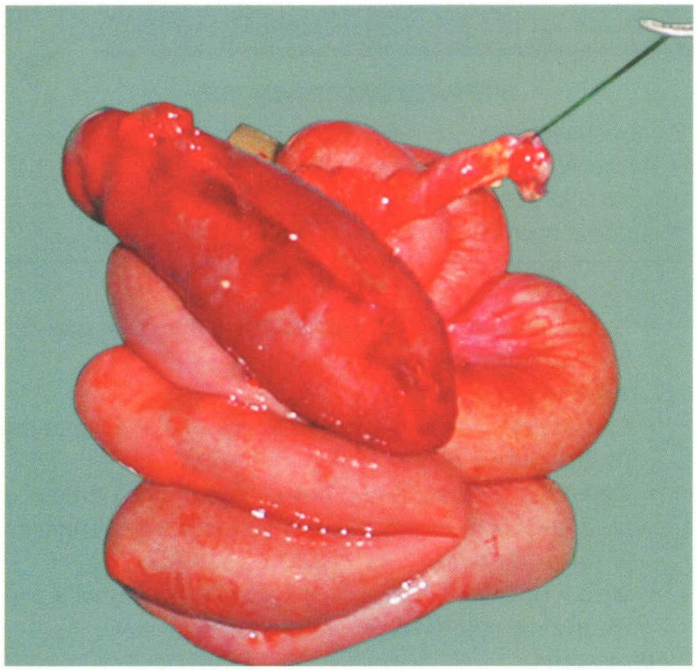


Fig. 30.90: Ileal atresia at surgery

DUODENAL ATRESIA (Key Box 30.32)

Types

Type 1: Complete atresia: It is the commonest atresia. The proximal dilated segment and distal collapsed segment are completely separated.

Types 2: There is no separation of the two parts. However, a **fibrous band** is in between.

Type 3: It is incomplete obstruction: There may be a web or stenosis.

Windsock deformity: An incomplete diaphragm, with central aperture (hole) with proximal dilatation

- *Anastomosis:* A special anastomosis named after Kimura is being described. It is a diamond shaped anastomosis between upper pouch (duodenum which is opened transversely and lower pouch (distal duodenum) which is opened longitudinally.
- Needless to mention here that general condition has to be taken care of with supportive therapy including fluid and electrolytes, total parenteral nutrition, nasogastric decompression and adequate urinary output.

KEY BOX 30.32

DUODENAL ATRESIA—ASSOCIATED LESIONS

- Annular pancreas, incomplete rotation of the gut
- Down's syndrome (trisomy 21)
- Maternal hydramnios
- Congenital heart disease
- Anorectal malformations

- Clinical features of obstruction manifest within 48–72 hour in the form of obstruction.
- Atresia means imperforation; stenosis means narrowing.
- Duodenal atresia presents as vomiting with or without bile minimal distension and visible gastric peristalsis.
- Jaundice
- **X-ray abdomen erect: Double-bubble** in duodena atresia.

Treatment

Duodenal atresia: Duodenojejunostomy by anastomosing dilated duodenum above the atresia to the jejunal loop.

SMALL INTESTINAL ATRESIA

Salient features

- It can affect jejunum (common) or ileum
- Like duodenal atresia, it can be associated with maternal hydramnios or malrotation of the gut.
- Exact reasons for atresia are not known. However, vascular variations in the mesentery such as V-shaped mesentery or due to occlusion of vessels in the intrauterine life are the possible factors.
- Like any obstruction, proximal bowel is dilated and distal bowel is collapsed. The colon is very small—microcolon.

GRIES FIELD MODIFICATION OF MARTIN'S CLASSIFICATION

- **Type I:** Simple stenosis. Mesentery is normal. It is mucosal atresia (Fig. 30.91).
- **Type II:** Proximal and distal bowel are connected by a fibrous band which has no lumen—atretic segment. Here again, mesentery is normal (Fig. 30.92).

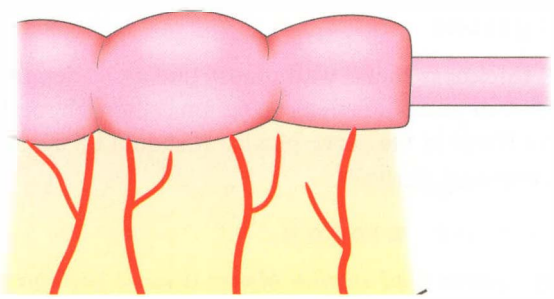


Fig. 30.91: Type I

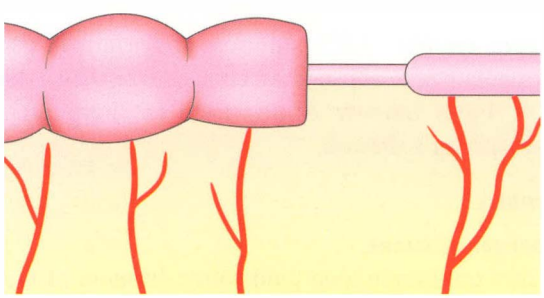
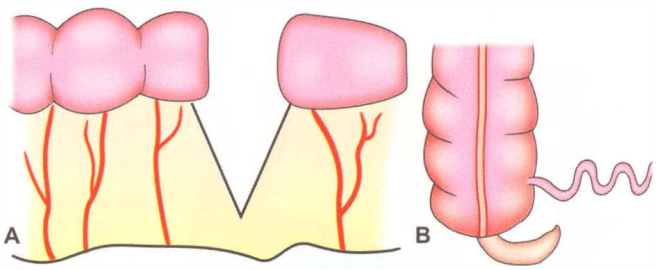


Fig. 30.92: Type II



Figs 30.93A and B: Type III

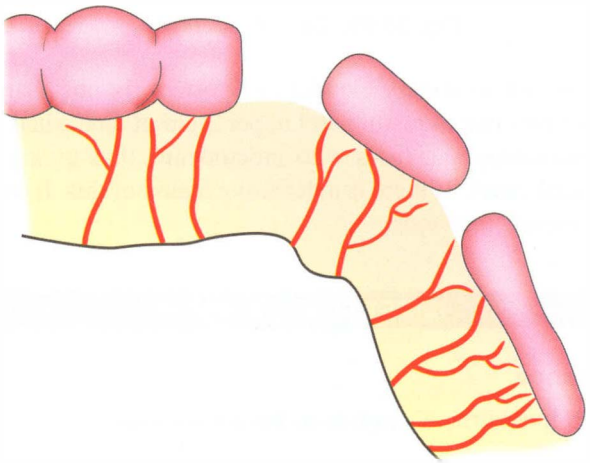


Fig. 30.94: Type IV

- **Type III:** Further divided into 2 types. IIIa refers to atresia with V-shaped loss of mesentery in between, IIIb refer to complete jejunal atresia with coiled ileum. It has been called Christmas tree deformity (Figs 30.93A and B).
- **Type IV:** Multiple atretic segments including mesentery (Fig. 30.94).

Please note: *Apple-peel* or *Christmas tree deformity*. Here, obstruction is usually in the proximal jejunum, which is supplied by the entire superior mesenteric artery (SMA). This results in a gap in the mesentery. So, rest of the small intestine is coiled around the ileocolic branch of the SMA.

Treatment

Resection, anastomosis—aim is to conserve as much as possible.

ARRESTED ROTATION WITH BANDS

- It is a congenital anomaly wherein the caecum and right colon are found on the left side (Fig. 30.95).
- As a result of this, there are peritoneal bands which run across from left to right and cause intestinal obstruction (duodenal obstruction).

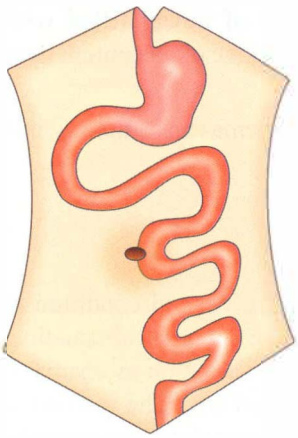


Fig. 30.95: Arrested rotation—malrotation

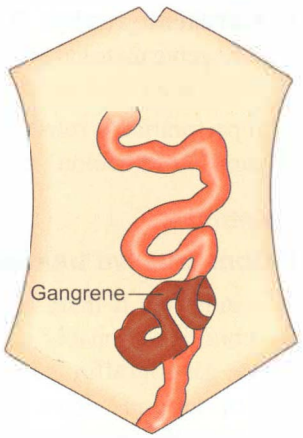


Fig. 30.96: Volvulus neonatorum

- One such band is called **transduodenal band of Ladd** which compresses the second part of duodenum and gives rise to obstruction (features similar to duodenal atresia).
- Differential diagnosis is duodenal atresia.

Treatment

Laparotomy, division of band and fixation of the caecum in the right iliac fossa.

VOLVULUS NEONATORUM

It is a complication of arrested rotation with bands which predisposes to midgut volvulus (small bowel). These unfortunate infants undergo massive resection of the bowel if it is gangrenous (Fig. 30.96). Such massive resections give rise to short gut syndrome. Thus, the patient becomes a digestive cripple. If the loop is not gangrenous, it is treated by laparotomy, untwisting of bowel and division of bands.

MECONIUM ILEUS

- It is a neonatal manifestation of **mucoviscidosis** of the pancreas wherein the mucus is thick and viscid. This, along with meconium, produces obstruction. **There may be ileal atresia**, which might have precipitated meconium ileus (Fig. 30.97). Majority of meconium ileus is coupled by complications such as **gangrene, perforation** and **peritonitis**. As a sequel to peritonitis, calcification and adhesive meconium obstruction develop.
- Infants present with abdominal distension, bilious vomiting and failure to pass meconium.
- Plain X-ray shows distended bowel and **mottling** due to calcification.
- **Soap bubble sign** or **Neuhauser's sign:** Ground glass appearance in the right lower quadrant due to viscid meconium mixed with air.

- **Ultrasonography:** Dilated loops of bowel filled with echogenic material are highly suggestive of meconium ileus rather than ileal atresia.
- If perforation is ruled out, barium enema can be done which shows microcolon.

Treatment

I. Conservative treatment

It is indicated if there is no peritonitis, general condition of the child is reasonably good or if there is partial obstruction. Dilute gastrograffin is introduced into the colon as enema. It fills up terminal ileum. It absorbs fluid from the interstitial space into the lumen because it is hyperosmolar. Consequently, the meconium becomes soft and it is rejected naturally. Hypervolaemia is to be corrected during this.

II. Nonresectional procedures

A. Bishop-Koop operation (Fig. 30.98)

- Ileum is divided in the proximal healthy part. This proximal ileum is anastomosed to the ascending colon to relieve obstruction—end-to-side anastomosis.
- Distal ileum containing thick meconium pellets is brought outside as a fistula and regular saline washes are given to dilute the meconium. Mucous fistula needs to be closed after a few weeks.

B. Santulli operation (Fig. 30.99)

- In this operation, proximal ileum is brought out as ileostomy. Distal ileum is anastomosed to proximal ileum as end-to-side anastomosis.

III. Resection

Resection is the surgery of choice today as more cases present with short segment obstruction. Long-term results and complications are much less than nonresectional procedures.

ANORECTAL ANOMALIES

Developmental anatomy

- To start with, there is a common chamber called cloaca, which is later divided into 2 chambers, anteriorly allantois gives rise to urinary bladder and posteriorly postallantoic gut gives rise to rectum and upper 2 cm of anal canal.
- Postallantoic gut fuses with proctodeum, thus giving rise to anal canal. If there is a defective fusion of this, it results in imperforate anus.

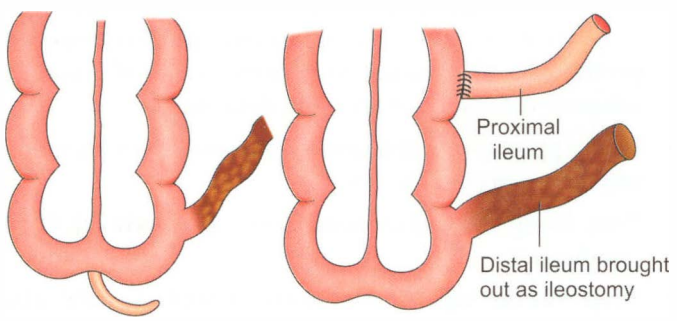


Fig. 30.97: Meconium ileus **Fig. 30.98:** Bishop-Koop operation

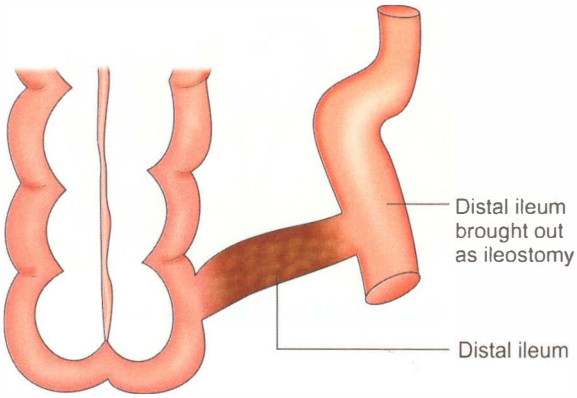


Fig. 30.99: Santulli operation

IMPERFORATE ANUS

Incidence

1:4500 live births. Common in female children.

Types of imperforate anus

I. Low anomalies: It refers to termination of the bowel below the anorectal bundle.

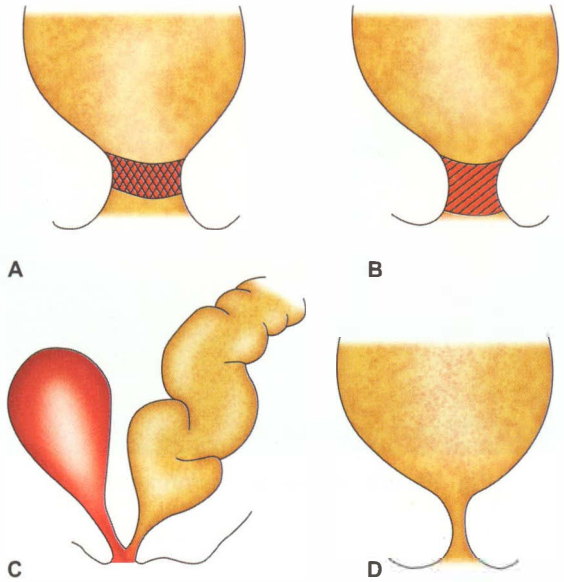
1. **Covered anus** (Fig. 30.100A)
   The anal orifice is covered by a tag of skin.
2. **Membranous anus** (Fig. 30.100B)
   Covered with a thin membrane
3. **Anterior ectopic anus** (Fig. 30.100C)
   Anus is situated anteriorly
4. **Stenosed anus** (Fig. 30.100D)
   Anal orifice is microscopic

II. High anomalies: They are supralevator—the bowel terminates above the anorectal bundle.

1. Anorectal and agenesis, with fistula: Rectovesical, rectovaginal, rectourethral fistulae (Fig. 30.101A).
2. Rectal atresia: Colon ends as a blind pouch below (Fig. 30.101B).
3. Cloaca: In this variety, bowel, urinary and genital tracts open into a common chamber. This occurs in females only.

Diagnosis—Wangensteen's invertogram

12 hours after birth, the child is held upside down (12 hours is the time for the gas shadow to reach the distal portion of the gut). A metal coin is strapped to the site of anus and X-ray is taken. If the gas shadow is above the pubococcygeal



Figs 30.100A to D: Low anorectal anomalies

line, it is a high anomaly. If the distance between the coin and gas shadow is more than 2.5 cm, it is a high anomaly. If the gas shadow is below the pubococcygeal line, it is a low anomaly. Pubococcygeal line is called as Stephen's line.

Treatment

I. **Low anomaly:** Easy to treat, division of membrane or skin followed by dilatation is all that is required with some amount of plastic reconstruction (anoplasty is necessary).

II. **High anomaly:** Repaired by 3-stage procedure:
- 1st stage: Preliminary transverse colostomy to relieve intestinal obstruction.
- 2nd stage: When the child is 8–10 kg of weight, a "pull-through" operation is done with division of fistula.
- 3rd stage: After 2 months, colostomy is closed.

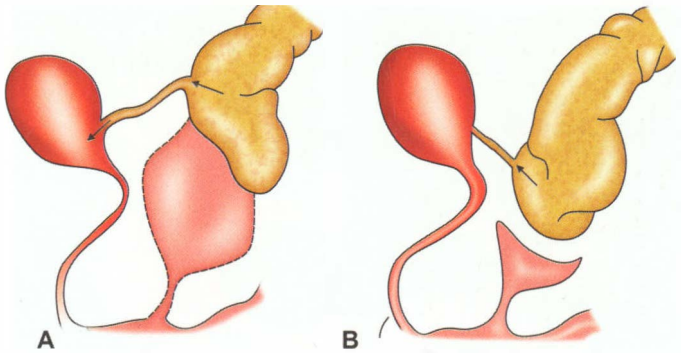
CAUSES OF INTESTINAL OBSTRUCTION AS PER AGE

Neonates: 0 to 7 days (Figs 30.102 to 30.109)

1. Atresia and stenosis
2. Hirschsprung's disease
3. Arrested rotation with bands
4. Volvulus neonatorum
5. Meconium ileus
6. Imperforate anus

Young patients: Up to 30 years

1. Obstructed hernia
2. Adhesions
3. Tuberculous stricture ileum
4. Crohn's disease of ileum
5. Tuberculous peritonitis with adhesions
6. Meckel's diverticulum with band
7. Adult intussusception



Figs 30.101A and B: High anorectal anomalies

Middle-aged patients: 30–60 years

1. Adhesions
2. Obstructed hernia
3. Carcinoma left colon
4. Diverticulosis with stricture left colon
5. Gall stone ileus
6. Sigmoid volvulus
7. Mesenteric vascular occlusion
8. Adult intussusception

FOOD BOLUS OBSTRUCTION

This complication can occur, particularly when a GJ or partial gastrectomy is done.

Factors precipitating this condition

- Unmasticated, undigested particles
- Coconut pieces, jackfruit seeds and gulped coins, etc.
- They get impacted in the terminal ileum which is the narrowest portion of the gut.

Treatment

Squeeze the bolus into the caecum. Otherwise, enterotomy and removal may be necessary.

OBSTRUCTION DUE TO INTERNAL HERNIA

Syn: Stammer's hernia

1. These are rare causes of intestinal obstruction. Due to some congenital defect in the mesentery, the floating, mobile intestines can herniate (Key Box 30.33). The defect may be in:
   - Mesentery
   - Transverse mesocolon or
   - Broad ligament
   - Foramen of Winslow

   Hence, whenever a surgical procedure is done for resection of the bowel or GJ, etc. Once the anastomosis is completed, the rent in the mesocolon as in GJ or rent in the small bowel mesentery should be closed.
2. Herniation can also occur through one of the potential spaces (fossae) in and around a viscus.

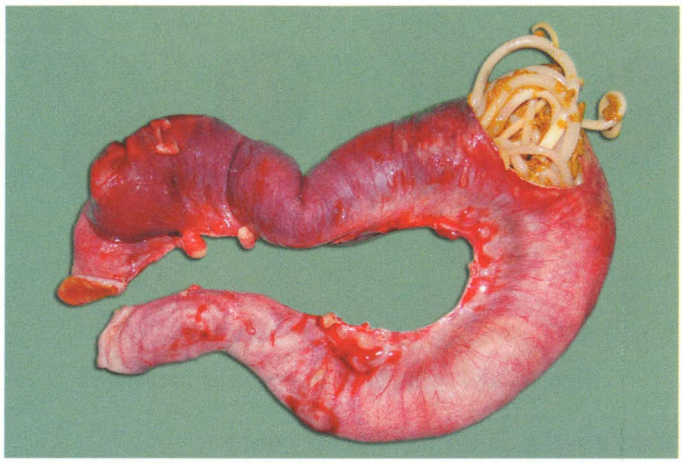


Fig. 30.102: Worm ball obstruction

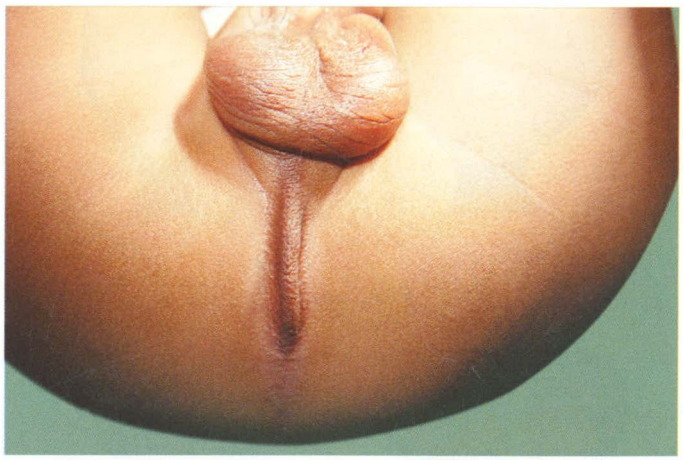


Fig. 30.103: Anorectal malformation

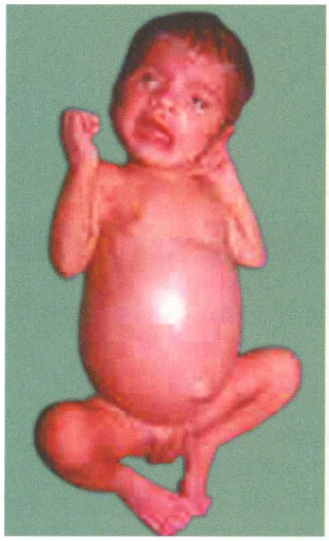


Fig. 30.104: Ileal obstruction

Fig. 30.105: Double-bubble appearance

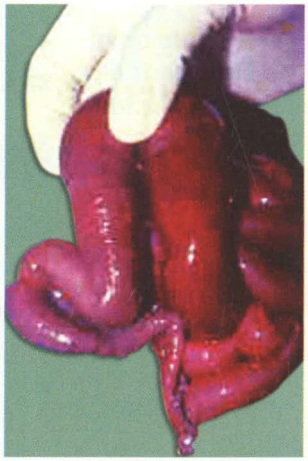


Fig. 30.106: Hirschsprung's disease

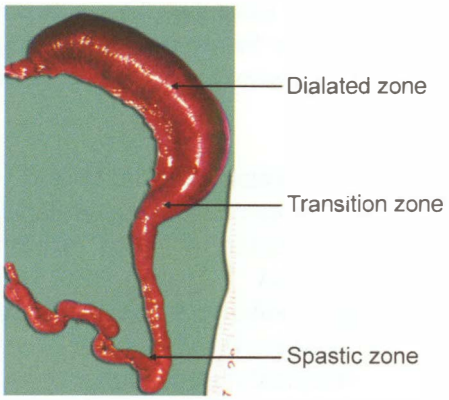


Fig. 30.107: Various segments of the large intestine in Hirschsprung's disease

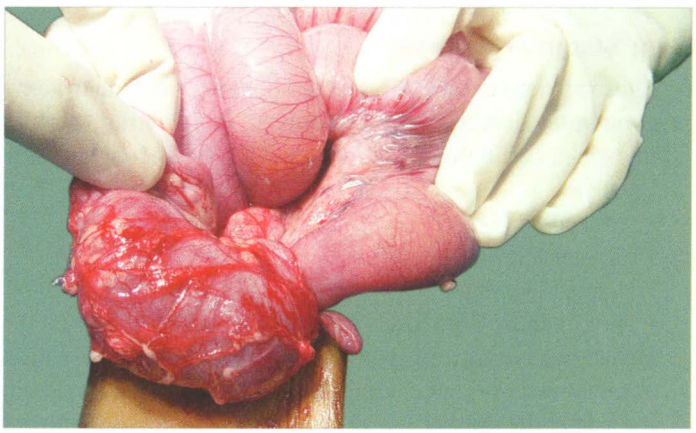


Fig. 30.108: Ileocaecal intussusception

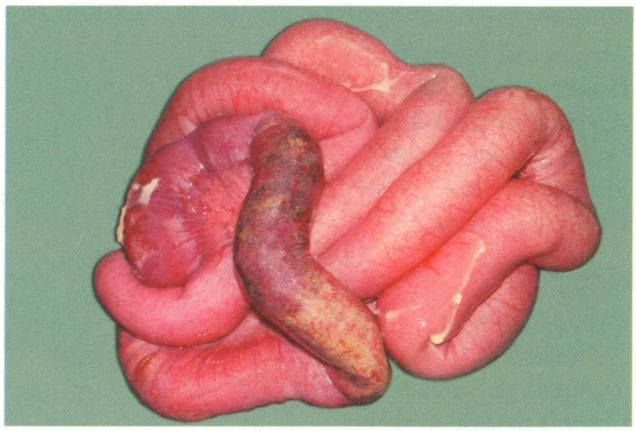


Fig. 30.109: Necrotising enterocolitis with patchy gangrene

(*Courtesy:* Professor Vijaykumar, Department of Paediatric Surgery, KMC, Manipal)

Duodenal fossa

- Left paraduodenal fossa: Inferior mesenteric vein lies very close to the free border here.
- Right duodenojejunal fossa: Superior mesenteric artery runs in its free border.

Colonic fossa

- Superior ileocaecal fossa
- Inferior ileocaecal fossa
- Sigmoid fossa

PEARLS OF WISDOM

These are the rare causes of intestinal obstruction to be kept in mind.

PARALYTIC ILEUS (NEUROGENIC ILEUS)

In this condition, there is a failure of transmission of parasympathetic impulses from one segment to the other. That means, there is a failure of parasympathetic mechanism, which results in paralysis of bowel. It gives rise to a large collection of fluid and gas within the bowel resulting in distension.

Causes

1. **Postoperative:** Exposure of the intestines, handling of the intestines, contamination with blood, foreign body, etc. causes temporary suppression of parasympathetic activity and results in paralytic ileus. This gets aggravated by allowing oral fluids too early.
2. **Following peritonitis**

Pus is the chief cause. Bile, blood, etc. are other causes. Bacterial toxins also prevent movement of the bowel.

3. **Reflex:** Following fracture spine, retroperitoneal haemorrhage, etc.
4. **Hypokalaemia:** K^+ is the chief intracellular ion of all cells including muscles. Hypokalaemia causes generalised muscle weakness including muscles of the bowel.

Clinical features

1. **Abdominal distension** is gross. Tympanitic note all over is a feature. Respiratory and cardiac functions are impaired.
2. **No colicky pain abdomen** (in dynamic obstruction, colicky pain is a feature). Dull pain occurs due to distension of the abdomen.
3. **Failure to pass flatus,** effortless vomiting is also characteristic of paralytic ileus.
4. On auscultation, **tinkling sounds** are heard due to shift of fluid from one coil of bowel to the other (Key Box 30.34).
5. Severe fluid, electrolyte and protein depletion occur.

KEY BOX 30.33

INTERNAL HERNIA

- Wherever a defect is present which is an anatomically present recesses or congenital or following surgery, loop of small intestine can herniate resulting in hernias.
- They are difficult to diagnose. The diagnosis is by exclusion
- Triggering factors: Inflammation in the vicinity with or without an adhesion and band trigger herniation and complications.
- Most of these cases present as acute abdomen with features of intestinal obstruction.
- CT scan is the best investigation in such cases
- It should be remembered that an important vessel runs in the close vicinity of these sites in most of the cases.
- Treatment is reduction, suturing the defect or resection anastomosis if the intestine is gangrenous taking care not to damage the vessel. Decompression without dividing the constricting ring may be required.

KEY BOX 30.34

THREE TYPES OF BOWEL SOUNDS

- Normal: Once in 20 seconds, low pitch
- Borborygmi: Loud, noisy, which occurs once in 5–10 seconds.
- Tinkling sounds: Mild metallic sounds as in paralytic ileus

Investigations

1. Plain X-ray abdomen erect demonstrates distended loops of bowel (Fig. 30.110).
2. Electrolyte study and correction of any abnormality.

Treatment

- Basic principle of treating paralytic ileus is **drip and suction**.
- The cause of paralytic ileus has to be treated first, e.g. if there is **hypokalaemia, supplement potassium**. If there is pus in the peritoneal cavity **drain** it.
- **Ryle's tube aspiration,** to give rest to the gut.
- Intravenous fluids, supplementation of ions, correction of dehydration, oliguria, etc. Such treatment is continued for 3–4 days.
- Ryle's tube is removed when abdomen is soft, bowel sounds are heard and patient has passed flatus. Clear oral fluids are started for 2–3 days followed by soft diet. Small bowel activity returns within 12–18 hours, followed by colon which starts functioning within 36–48 hours. However, gastric functions may return ranging from 18 hours to 4 days.

PSEUDOINTESTINAL OBSTRUCTION

Acute colonic pseudo-obstruction (ACPO) is also called **Ogilvie's syndrome**. It is massive colonic distension in the absence of a mechanically obstructing lesion.

Fig. 30.110: Plain X-ray abdomen showing dilated bowel loops

KEY BOX 30.35

CAUSES OF COLONIC PSEUDO-OBSTRUCTION

1. Retroperitoneal irritation
   - Blood
   - Urine
   - Fracture spine and pelvis
2. Drugs
   - Levodopa
   - Tricyclic antidepressants
3. Metabolic
   - Uraemia
   - Diabetes
   - Myxoedema
   - Hypokalaemia
4. Viral infections

Pathogenesis

It occurs mainly due to **malfunctioning of sacral parasympathetic nerves (S2–S4)**. It results in atony of the descending colon resulting in functional obstruction. It is interesting to note that the **junction of the dilated and collapsed bowel is near the splenic flexure. This is the place wherein parasympathetic supply by vagus ends and sacral autonomic nervous system starts**. An increased sympathetic tone results in colonic dilatation due to inhibition of contraction (Key Box 30.35).

Clinical features

- Elderly bedridden patient with cardiac/lower respiratory illness are the victims. Aerophagia and drugs which decrease colonic mobility are precipitating factors.
- Failure to pass faeces and flatus for several days.
- Tachypnoea due to elevation of the diaphragm due to distended colon is common.
- Rectal examination reveals some faeces (in cases of mechanical obstruction, rectum is empty).
- Plain X-ray abdomen erect may or may not show one or two air fluid levels. Distension is mainly colonic.
- Carcinoma colon is to be differentiated by barium enema.
- Caecal perforation is a dangerous complication. Hence, look for right iliac fossa of tenderness.

Treatment

- It is conservative, provided acute abdomen is ruled out.
- Colonoscopic decompression is the method of choice.
- Prokinetic drugs such as cisapride or mosapride have been tried in selected cases.
- Rarely, even after colonoscopic decompression, caecal tenderness continues. If distension persists, laparotomy followed by tube caecostomy may have to be done.

INTESTINAL OBSTRUCTION—SPECIAL CAUSES

1. Adhesive obstruction

- Firstly, strangulation should be ruled out by clinical and radiological tests. **Extended nonoperative therapy** may be advised, e.g. if one can wait for 48 hours in a case of intestinal obstruction. In these cases 4–6 days of waiting is sometimes worth, especially in a patient who has been operated many times earlier.
- During this extended period, careful monitoring is important to look for any new symptoms/signs of strangulation.
- **Early postoperative adhesions** (bread and butter adhesions) can also be given an extended nonoperative therapy because mostly it is **partial obstruction.**

2. Intestinal obstruction in Crohn's disease

- As far as possible, **resection should be avoided** in Crohn's disease because the aim is to save as much as possible.
- 30% of patients eventually develop obstruction which requires resection.
- Again, if possible do **strictureplasty**
- Strictureplasty should not be done in patients with intra-abdominal abscesses or intestinal fistulae.
- Some cases of Crohn's obstruction also **respond well to medications—one more reason for nonoperative treatment.**

3. Intestinal obstruction in pregnancy

- The commonest cause of intestinal obstruction in pregnancy and puerperium is **adhesive bands**.
- **Volvulus** is the second commonest cause of intestinal obstruction—intestine volvulates around an adhesive band

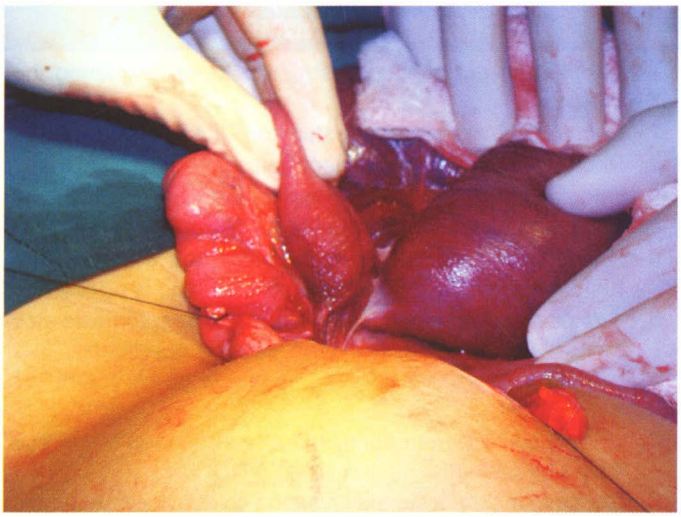


Fig. 30.111: Pregnant lady with adhesions causing intestinal obstruction

and that is why the primary cause is adhesive bands (Fig. 30.111).

- **Inguinal hernia** and **intussusception** are the other causes.
- The consequences of intestinal obstruction in pregnancy **do not differ** from those found in nonpregnant women except that in the pregnant patient, a second entity, the foetus, is also threatened.
- Most of the intestinal obstructions tend to occur in the **third trimester**.
- Maternal **mortality** for intestinal obstruction in pregnancy (10–33%) is **higher** than in nonpregnant patients.
- In the first half of the pregnancy, nausea, vomiting and episodes of constipation are quite common. Hence, they are confused for hyperemesis gravidarum, acute duodenal ulcer and gastritis.
- **In the second half of the pregnancy,** symptoms can be confused for toxaemia, constipation, Braxton Hicks contractions, etc.
- **Ultrasound** can help in the diagnosis by detecting dilated intestinal loops, fluid in the peritoneal cavity and intussusception. It also helps to rule out ovarian torsion, gall stone disease, etc.
- **Premature labour can be prevented with tocolysis (abolition of uterine contraction).**
- **Abdominal surgery in the third trimester does not induce labour.**
- Negative laparotomy carries a small risk of disturbing the pregnancy.
- **Volvulus and intussusception** are the two major causes of small intestinal obstruction in pregnancy. Both can give rise to gangrene. Hence an intervention is required before gangrene sets in.
- ***The diagnosis and treatment of a pregnant patient suspected of having a bowel obstruction should be no different from those given to a nonpregnant one.***

4. Ileosigmoid knotting

It is rare type of knotting between sigmoid and ileum carrying significant mortality if not treated timely (Key Box 30.36).

5. Iliac crest graft hernia

- This is also a rare hernia following bone graft removal from the iliac crest for treatment of fractures.

KEY BOX 30.36

ILEOSIGMOID KNOTTING

- It is also called compound volvulus.
- Predisposing factor is long pelvic mesocolon.
- The ileum twists around the sigmoid colon resulting in gangrene of ileum or sigmoid colon or both.
- Even though sigmoid is also twisted, features are similar to small intestinal obstruction—not massive distension as seen in sigmoid volvulus.
- Resection anastomosis of gangrenous segment followed by anastomosis of the bowel (ileoileal/colic and colocolic).

- Iliac crest is the common donor site of bone graft. If the bone graft removed is large, the intestines can herniate resulting in intestinal obstruction.
- Tenderness in the surgical site scar can cause diagnostic difficulty.
- It is confused for haematoma
- CT scan gives the diagnosis

Following is the case report of a patient who had undergone iliac crest bone graft and developed hernia (Figs 30.112 to 30.114).

6. Acute large bowel obstruction

Details about the causes of large bowel obstruction, pathophysiology and the treatment is given in Chapter 29. However a few important points have been given in ten commandments.

CLINICAL NOTES

A 50-year-old man presented to the casualty with abdominal pain, vomiting and distension. Features were suggestive of intestinal obstruction. On examination, there was a buldge in the region of right iliac fossa—more lateral. There was a scar of bone graft incision. On questioning the patient he says that he had fracture humerus. Two months back, nailing and bone grafting had been done. Ultrasound revealed bowel loops. CT scan showed a large bone graft defect with herniation of intestines resulting in intestinal obstruction. Emergency surgery, reduction of hernia contents and mesh repair was done. Recovery was uneventful.

Fig. 30.112: Iliac bone graft site hernia

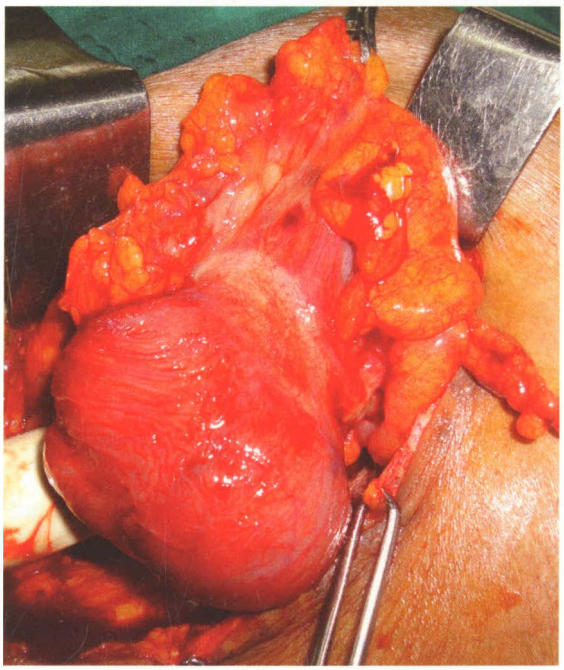


Fig. 30.113: Caecal herniation

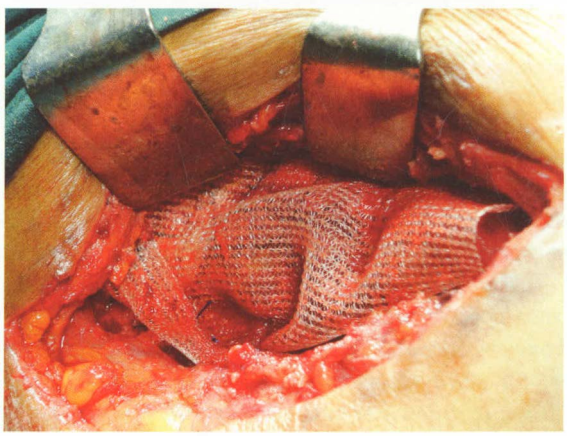


Fig. 30.114: Caecal hernia through iliac bone graft defect—mesh repair

TEN COMMANDMENTS

1. Should rule out pseudo-obstruction before exploring the abdomen.
2. Should do limited contrast study or CT scan
3. Should resuscitate the patient before surgery
4. Should take into account, the general condition of the patient before resection anastomosis.
5. Should consider right hemicolectomy or extended hemicolectomy for right-sided growth which is operable.
6. Should consider single stage resection anastomosis also for left-sided growth provided general condition of the patient is good and on table lavage is given before the anastomosis.
7. Should consider exteriorisation of gangrenous bowel in a very sick patient.
8. Should consider a simple diversion colostomy in moribund patients in rectosigmoid obstructions (it is a common problem).
9. Should not do anastomosis in cases with faecal contamination, peritonitis, haemodynamic instability or possible ischaemia of the remaining colonic segments.
10. Should mark the probable stoma site

MALROTATION AND MIDGUT VOLVULUS

Introduction

- The incidence of malrotation is 1 in 500 infants.
- The male to female ratio is 2 : 1.
- Malrotation with midgut volvulus may become rapidly life-threatening. The previously healthy infant with bilious vomiting is a characteristic presentation of malrotation. Usually it presents with bilious vomiting, failure to thrive and features of intestinal obstruction (Fig. 30.115).
- In approximately 60% of patients, malrotation presents by one month of age. Another 20–30% of patients present at age 1–12 months. Thereafter, it can present at any age, and is seen in adults and even the elderly.

Basic pathophysiology

- A volvulus is a complete **twisting of a loop of intestine around its mesenteric attachment site.** This can occur at various locations of the GI tract, including stomach, small intestine, caecum, transverse colon, and sigmoid colon. Midgut malrotation refers to twisting of the entire midgut about the axis of the superior mesenteric artery (SMA).
- **Malrotation is any deviation from the normal 270° counterclockwise rotation** of the bowel that occurs during embryogenesis (Figs 30.115 and 30.116).
- At the fourth week of gestation, the gastrointestinal system is a straight tube centrally located in the abdomen. During the ensuing 8 weeks, the midgut rotates and becomes fixed to the posterior abdominal wall. **Arrest of development at any stage narrows the mesenteric base and impairs fixation, leaving the bowel at high risk for volvulus.**

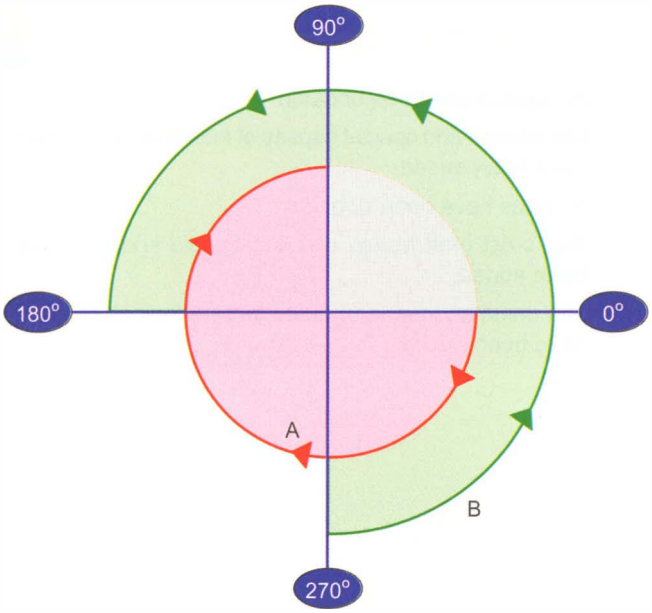


Fig. 30.115: Midgut volvulus: (A) Clockwise rotation resulting in superior mesenteric ischaemia shown in red line, (B) Anticlockwise untwisting to be done at surgery is shown as green line

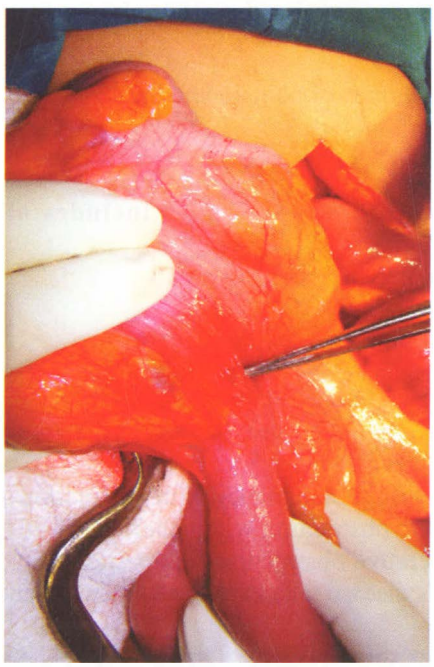


Fig. 30.116: Malrotation with band

- The resultant shortened mesenteric pedicle predisposes to midgut volvulus, a clockwise rotation around the superior mesenteric artery axis that can lead to bowel ischaemia.

Investigations

- Conventional radiographs are neither sensitive nor specific for malrotation.
- On the upper GI series, it is crucial to locate the position of the duodenojejunal junction (DJJ). The DJJ must be at least over (but more reassuringly lateral to) the left vertebral pedicle and at the same height as the duodenal bulb on a well-centred view. If the DJJ does not meet these two criteria, malrotation is diagnosed.
- **Signs of midgut volvulus include an abrupt termination, or break of the contrast column and the corkscrew (apple peel, or barber pole) sign.**
- On ultrasound (US) and computed tomography (CT), the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) relationship may be reversed.
- Normally, the SMV is to the right of the SMA; with malrotation, the SMV may occupy a position directly anterior or to the left of the SMA.

Treatment

- **The Ladd procedure** remains the cornerstone of surgical treatment for malrotation today.
- ***A classic Ladd procedure*** is described as reduction of volvulus (if present), division of mesenteric bands, **placement of small bowel on the right and large bowel on the left of the abdomen, and appendicectomy.**
- A laparoscopic variation of the Ladd procedure has been used in some centres, with the general advantage of decreased adhesions and scarring, but good visualisation of the entire bowel is necessary. Laparoscopy is a particularly suitable investigative procedure for children presenting with acute abdomen.

'Clues' to the causes of intestinal obstruction are mentioned in Key Box 30.37.

KEY BOX 30.37

INTESTINAL OBSTRUCTION

'Clues' to the causes of obstruction

• Operative scar	– Adhesions
• Groin swelling	– Obstructed hernia
• Gross distended intestinal loop	– Sigmoid volvulus
• Step ladder peristalsis	– Small intestinal obstruction
• Red currant jelly stools	– Intussusception
• Deformed umbilicus in adult child	– Meckel's diverticulum with band
• Melanosis of lips, mucosa	– Intussusception in adults (PJ syndrome)
• H/o constipation/bleeding per rectum	– Colonic obstruction (cancer)
• Emaciated, young patient with loose stools/weight loss/fever	– Ileocaecal tuberculosis TB stricture
• Elderly, hypertensive severe abdominal pain, tachycardia, tachypnoea, acidotic breathers	– Mesenteric vascular occlusion

No wonder 'a surgeon should have an eagle eye'!

INTERESTING 'MOST COMMON' IN INTESTINAL OBSTRUCTION

- Most commonly encountered disorder of intestines is intestinal obstruction.
- Most common cause of small intestinal obstruction is intra-abdominal adhesion.
- Most commonly performed, simple diagnostic investigation in case of obstruction is plain X-ray abdomen in erect position.
- Most common abdominal symptom of intestinal obstruction is colicky abdominal pain.
- Most commmon congenital anomaly of small intestine is Meckel's diverticulum.
- Most common presentation of Meckel's diverticulum in children is bleeding.
- Most common cause of intestinal obstruction in infants between 6 and 18 months is intussusception.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated
- Malrotation and special causes of intestinal obstruction have been added.
- Atresias have been updated
- Iliac crest graft hernia and ileosigmoid knotting have been added.
- Ten commandments of acute large bowel obstruction have been added.

MULTIPLE CHOICE QUESTIONS

1. Which of the following is true for closed loop obstruction?
A. Can occur with constrictive growth in the hepatic flexure
B. Ileocaecal valve is incompetent
C. Perforation of the sigmoid colon is common
D. Occurs with partial obstruction

2. Which of the following is not the cause of gangrene in intestinal obstruction?
A. Ileocaecal tuberculosis
B. Mesenteric vascular occlusion
C. Necrotising enterocolitis
D. Volvulus

3. Faeculent vomiting is pathognomonic of:
A. Jejunal obstruction
B. Terminal ileal obstruction
C. Duodenal obstruction
D. Colonic obstruction

4. Cardinal features of intestinal obstruction include all of the following *except*:
A. Colicky abdominal pain
B. Vomiting
C. Diarrhoea
D. Abdominal distension

5. The following is true in a plain X-ray of abdomen in intestinal obstruction:
A. Caecum can appear as round shadow
B. Ileum has valvulae conniventes
C. Colon has haustrations
D. Sigmoid appears shapeless

6. Features of strangulation includes all of the following *except*:
A. Tachycardia
B. Disappearance of pain abdomen
C. Fever
D. Acidosis

7. The features of viable bowel includes all of the following *except*:
A. Normal peristalsis
B. Normal pulsations are visible
C. Normal pink colour is present
D. Peritoneal sheen is absent

8. Conservative treatment is advocated in intestinal obstruction when there is:
A. Disseminated malignancy with obstruction
B. Complete obstruction with adhesions
C. Postoperative obstruction with peritonitis
D. Crohn's disease unresponsive to medications

9. 'Bent inner tube design', 'Omega sign', 'Bird's beak design' are all seen in:
A. Sigmoid volvulus
B. Caecal volvulus
C. Meckel's diverticulum
D. Bascule

10. The most common cause of intestinal obstruction in infants aged 6–18 months is:
A. Worms
B. Bands
C. Intussusception
D. Adhesions

11. Red currant jelly stools are characteristic of:
A. Worms B. Bands
C. Intussusception D. Adhesions

12. Dance's sign (*signe de dance*) is a feature of:
A. Worms B. Bands
C. Intussusception D. Adhesions

13. The investigation of choice in mesenteric vascular occlusion is:
A. Ultrasound abdomen
B. Plain X-ray abdomen
C. CT with or without angiogram
D. MRI

14. The most common cause of intestinal obstruction in neonates is:
A. Bands
B. Duodenal atresia
C. Imperforate anus
D. Meconium ileus

15. The most common congenital anomaly of small intestine is:
A. Bands
B. Duodenal atresia
C. Stenosis
D. Meckel's diverticulum

16. Features of paralytic ileus include the following *except*:
A. Gross abdominal distension
B. Pain abdomen
C. Failure to pass flatus
D. Tinkling sounds

17. Melanosis of lips and mucosa with intestinal obstruction should arouse the suspicion of:
A. Gardner's syndrome
B. Turcot's syndrome
C. Peutz-Jeghers syndrome
D. Down's syndrome

18. Which is the factor precipitate sigmoid volvulus?
A. Short colon
B. Broad attachment at the base
C. Empty colon
D. Long mesentery of the colon

19. Common factor precipitating sigmoid volvulus in patients with parkinsonism, multiple sclerosis, hypothyroidism is:
A. Diarrhoea
B. Constipation
C. Drugs
D. Long mesentery of the colon

20. Investigation of choice for detecting bleeding Meckel's diverticulum is:
A. CT scan
B. Pet scan
C. MRI scan
D. Technetium scan

21. Presence of intramural air is diagnostic of:
A. Gall stone ileus
B. Sigmoid perforation
C. Duodenal atresia
D. Intestinal gangrene

22. Following complications can occur after gastro-jejunostomy *except*:
A. Dumping syndrome
B. Intussusception
C. Volvulus
D. Stomal ulcer

23. Following are the causes of adult intussusception *except*:
A. Meckel's diverticulum
B. Submucous lipoma
C. Carcinoma caecum
D. Hypertrophy of Peyer's patches

24. The most common anomaly associated with Hirschsprung's disease is:
A. Down's syndrome
B. Hypothyroidism
C. Meckel's diverticulum
D. Anorectal atresia

25. Following are causes of paralytic ileus *except*:
A. Anastomotic leak
B. Retroperitoneal irritation
C. Hyperkalaemia
D. Fracture spine

ANSWERS

1 A	2 A	3 B	4 C	5 A	6 B	7 D	8 A	9 A	10 C
11 C	12 C	13 C	14 B	15 D	16 B	17 C	18 A	19 B	20 D
21 D	22 B	23 D	24 A	25 C					

31

Rectum and Anal Canal

- Surgical anatomy
- Carcinoma rectum
- Prolapse rectum
- Surgical anatomy of anal canal
- Haemorrhoids
- Anorectal abscess
- Fistula *in ano*
- Fissure *in ano*
- *VAAFT*
- Pilonidal sinus
- Sacrococcygeal teratoma
- Malignant tumours of anal canal
- Stricture of anal canal and rectum
- Anal incontinence
- What is new?/Recent advances

SURGICAL ANATOMY OF THE RECTUM

The rectum starts at the rectosigmoid junction, opposite the **third piece of sacrum**. It descends in the sacral hollow, passes through the pelvic floor, and ends in the anorectal junction, which is about 4 cm away from the anal verge. Anorectal junction is enclosed by puborectalis muscle posteriorly and in the lateral aspects. **The rectum is 12–15 cm in length** (Key Box 31.1).

Peritoneal covering

- Upper one-third is **completely covered** by peritoneum (> 11 cm from anal verge) (Figs 31.1 and 31.2)
- Middle one-third is **covered in front and lateral aspects** (6–11 cm).
- Lower one-third (0–6 cm) has **no extraperitoneal covering** but has two fascial condensation layers. Posteriorly, the strong **Waldeyer's** layer separates the rectum from lower sacral pieces and coccyx. At surgery, **stripping of this fascia results in uncontrollable bleeding from sacral plexus of veins,** which is underneath the Waldeyer's fascia.
- Anteriorly, the weak **Denonvillier's** fascia separates the rectum from prostate and bladder. **Stripping of this fascia results in troublesome bleeding from prostatic venous plexus.**
- Rectum is attached to side wall of pelvis by **lateral ligaments**, which contain middle haemorrhoidal vessels. These need ligation or coagulation during mobilisation of lower rectum.
- **Valves of Houston:** Despite the name rectum means straight, it is **never straight in adults.** It has one convexity on the left and two convexities on the right side. There are 3 **valves of Houston** (prominent mucosal folds), two on the left and one on the right.
- That portion of the rectum resting on the pelvic floor is called **ampulla**—dilated portion of the mid rectum.

Arterial supply

1. **Superior haemorrhoidal artery** is a branch of the superior rectal artery which is the continuation of the inferior mesenteric artery. It divides into right and left branches. The right branch divides into anterior and posterior branches which supply the rectum (Fig. 31.3).

KEY BOX 31.1

SOME INTERESTING FEATURES OF RECTUM

1. Rectum means straight but it is not
2. Coverings of the peritoneum different in different levels
3. Eventhough it is a part of the large intestine, taenia, appendices epiploicae and sacculations are absent.
4. Middle curve marks the anterior peritoneal reflection. It is about 12–15 cm above anus.
5. Rectal carcinoma has high recurrence because of lack of serosal layer and close relation to other pelvic viscera.
6. Principal route of lymphatic drainage is upwards towards para-aortic nodes.

KEY BOX 31.2

RECTOVESICAL POUCH—RECTOUTERINE POUCH

- After investing the upper rectum, pelvic peritoneum is reflected anteriorly in males urinary bladder—thus form rectovesical pouch and in females it reflects onto uterus and thus form rectouterine pouch.
- It is one of the sites of transcoelomic spread of malignant cells.
- Maligant cells settle down in this most dependent part of the abdominal cavity and grow.
- They are palpable by rectal examination, a shelf like feeling—popularly called Blumer's shelf.
- Thus, if per rectal or per vaginal examination findings suggest presence of Blumer's shelf—it means hard deposits are felt and the case is inoperable.
- Rectovesical pouch is also the site of pelvic abscess
- It is diagnosed by per rectal or per vaginal examination
- Pus can be drained through rectum or through posterior fornix.
- Aspiration of blood from rectovesical pouch through posterior fornix also indicate intraperitoneal bleeding—may be ruptured ectopic.

2. **Middle haemorrhoidal artery,** a branch of internal iliac artery, runs in the lateral ligament of the rectum.

3. **Inferior haemorrhoidal artery,** a branch of internal pudendal artery supplies the lower rectum.

Venous return (Fig. 31.4)

The rich submucous plexus of veins surrounding the ampulla forms external rectal plexus. The venous drainage from here flows in two directions.

1. **Upwards** to drain into **superior rectal veins.** These join inferior mesenteric veins, which in turn drain into the portal system.

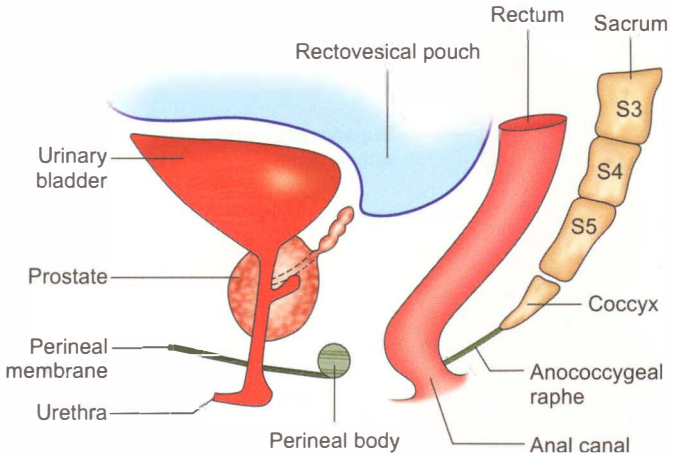


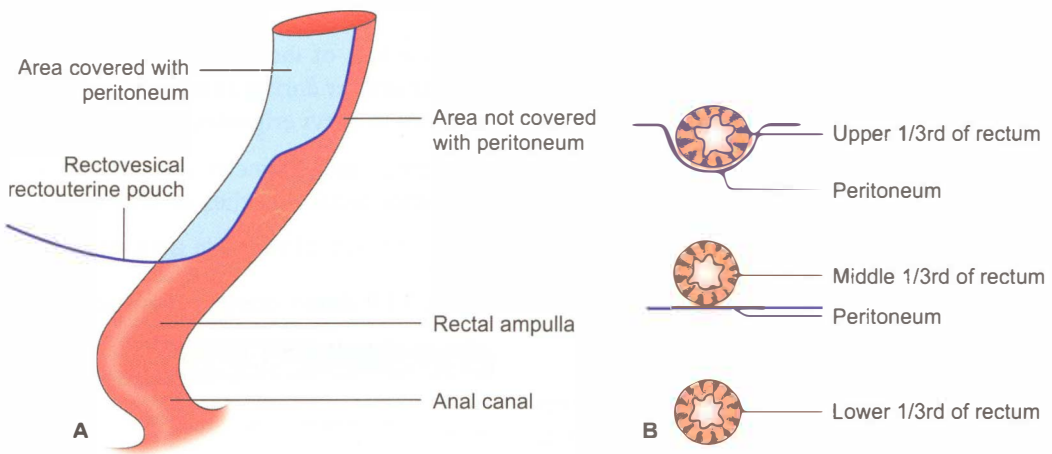
Fig. 31.1: Sagittal section through the male pelvis showing the location of the rectum and some of its anterior and posterior relations

2. **Across** to drain into middle rectal veins, which run in the lateral ligament of the rectum along with middle rectal artery. Hence, the lateral ligaments have to be ligated and divided during resection of rectum.

   These veins drain into internal iliac veins (systemic circulation). Hence, rectum is a site of portosystemic anastomosis.

Lymphatic drainage of rectum (Fig. 31.5)

- **Upper 1/3rd of rectum** is completely enclosed by peritoneum and the **middle l/3rd of rectum** is covered in front and on the sides by peritoneum. From these areas, lymphatic drainage always occurs in the **upward direction,** first to **(A)** pararectal nodes of Gerota followed by superior haemorrhoidal nodes, middle haemorrhoidal nodes and nodes at the origin of inferior mesenteric artery.
- From **lower l/3rd of rectum**, lymphatics spread in the **lateral direction** and can involve **(B)** internal iliac nodes.



Figs 31.2A and B: Peritoneal relations of the rectum

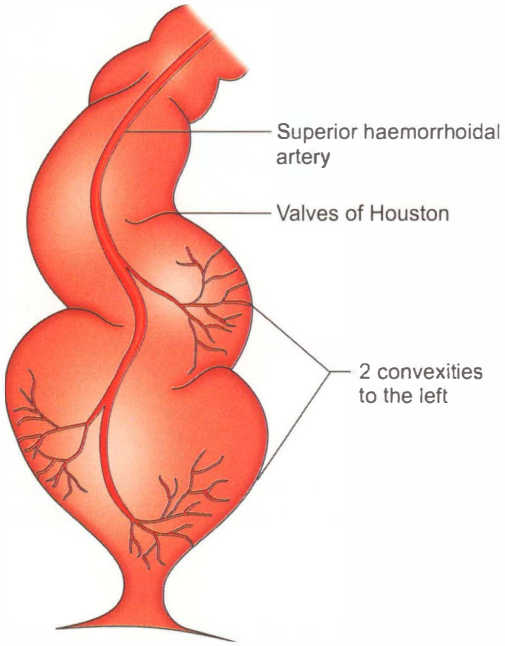


Fig. 31.3: Arterial supply of rectum

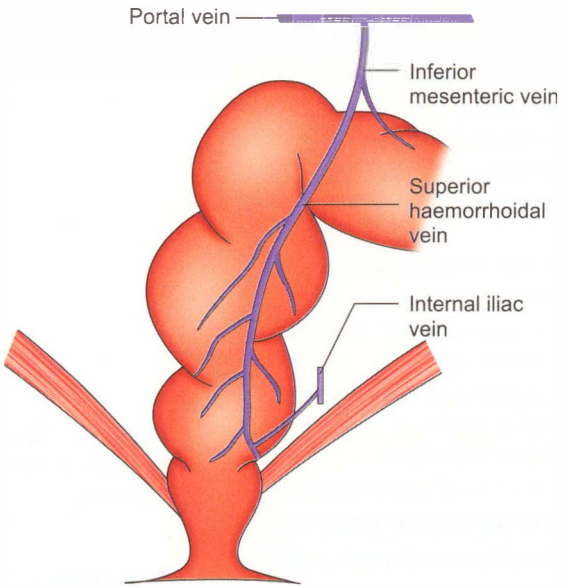


Fig. 31.4: Venous return

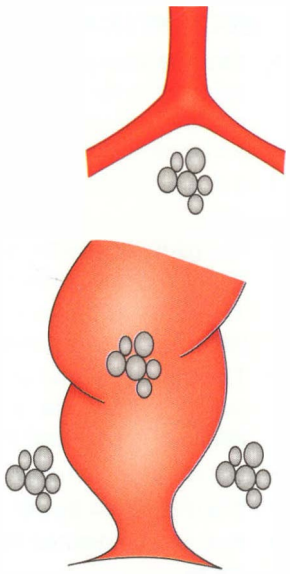


Fig. 31.5: Lymphatic drainage

Fig. 31.6: Digital rectal examination

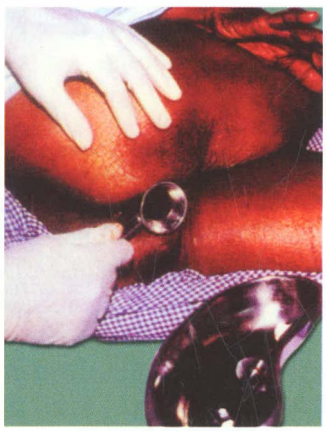


Fig. 31.7A: Proctoscopy

Fig. 31.7B: Sigmoidoscopy

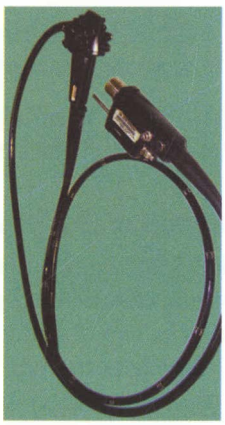


Fig. 31.8: Colonoscopy

- Lymph nodes are also present in the hollow of sacrum along median sacral artery **(C)**.

Lymphatics are present in the muscularis mucosa.

Nerve supply

- **Sympathetic:** The fibres come from hypogastric plexus, which is located at the aortic bifurcation at the level of L_5. Injury to this can cause absence of erection or dry orgasm. Fibres also come along with inferior mesenteric artery and superior rectal artery.
- **Parasympathetic:** (S2, S3, S4) by means of nervi erigentes from the hypogastric plexus and supply motor fibres to detrusor. Pain and ability to distinguish flatus and faeces is because of these fibres. Loss of mucosa of the rectum results in the loss of these sensations. During division of lateral ligaments or during anterior dissection of the bladder base, injury to nervi erigentes can occur.
- External anal sphincter and puborectalis are innervated by inferior rectal branches of internal pudendal nerve.

Examination of rectum and anal canal (Table 31.1)

Figure 31.9 shows common diseases of ano rectum.

CARCINOMA RECTUM

Aetiopathogenesis

Similar to carcinoma colon. However, precancerous conditions and risk factors are given as follows.

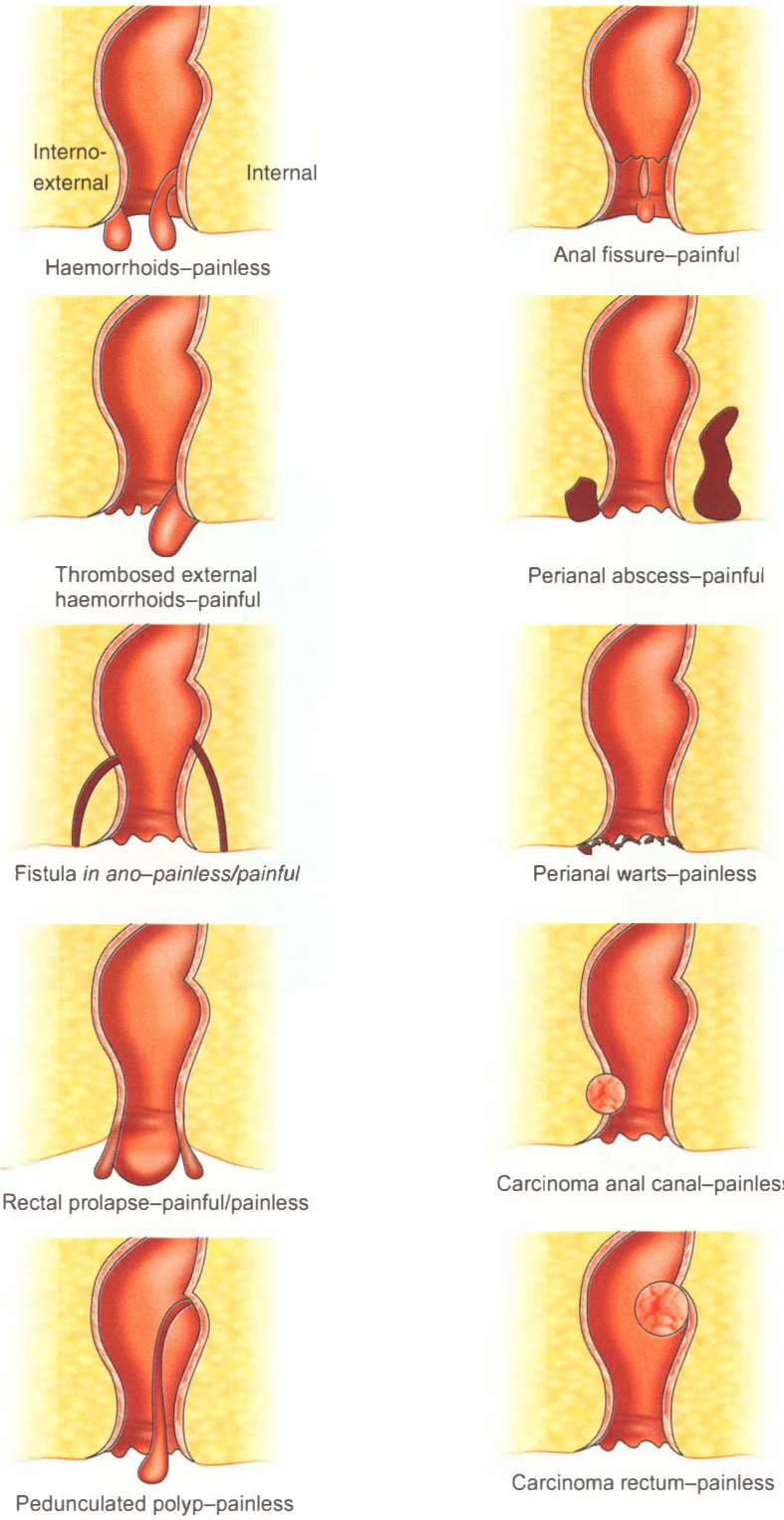


Fig. 31.9: Common diseases of ano rectum

These are the common anal/perianal conditions which can be diagnosed by inspection (only), or palpation or per rectal examination/ proctoscopic examination

Table 31.1 Examination of rectum and anal canal

Digital rectal examination (Fig. 31.6)	**Proctoscopy (Kelly's)** (Fig. 31.7)	**Flexible sigmoidoscopy** (Fig. 31.8)
• Lubricate the finger with lignocaine jelly • **Sim's position: Left lateral** is ideal position • Explain to the patient what you plan to do • Gently apply pressure on the external sphincter (anal opening) and slowly introduce the finger • Polyps, carcinomatous growths, strictures, thrombosed piles can be felt • Poor sphincter tone as in prolapse rectum, spinal cord paralysis	• Do a per rectal examination and rule out painful condition (*Fissure in ano*) • Proctoscope with obturator is introduced to full length • Piles are seen bulging into lumen as **obturator is withdrawn** • Biopsy from growth can be taken • Piles can be injected with sclerosants • Pelvic abscess can also be drained	• Flexible sigmoidoscope is 60 cm long • An enema is given before procedure • Growth, ulcers, bleeding diverticulae, polyps, colitis can be diagnosed and biopsy can be taken • Do not force instrument—it may perforate colon • It may deflate and derotate sigmoid volvulus

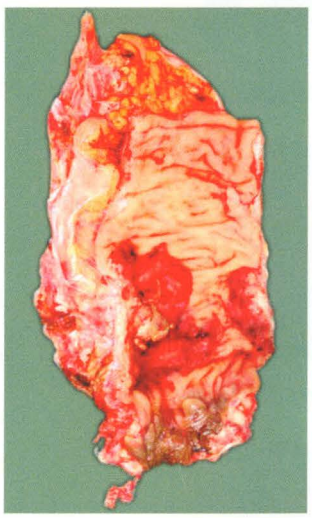


Fig. 31.10: Annular constricting lesion—patient presents with colonic obstruction

Fig. 31.11: Fibreoptic sigmoidoscopy showing growth in the upper rectum—biopsy proved signet ring carcinoma

Precancerous conditions

- Polyps in FAP, villous adenoma (*see* page 704)
- Ulcerative colitis (*see* page 677)
- Crohn's disease (*see* page 683)

Risk factors

- **S**moking, **A**lcohol, **D**iet (*see* page 709)
- Genetic
- Colorectal family (*see* page 705, 706)

Pathological types

1. **Annular** variety is common at the **rectosigmoid** junction. It presents with constipation and intestinal obstruction. It takes about a year for the growth to completely encircle the lumen of the gut (napkin ring deformity) (Fig. 31.10).
2. **Polypoidal** lesions are common in the **ampulla** of the rectum (Fig. 31.11).
3. **Ulcerative** lesions can occur anywhere in the rectum with **raised edges** and the growth occurs in the transverse direction.
4. **Diffuse** variety is similar to linitis plastica. It develops from ulcerative colitis. It has a poor prognosis.
5. **Colloid** variety is rare. The tumour contents are **gelatinous** due to increased mucus production. This variety is seen in young patients. The cell is filled with mucus and nucleus is displaced. It is called **'Signet Ring' carcinoma**. It is associated with poor prognosis (Fig. 31.12).

Clinical features of carcinoma rectum

- **Constipation** requiring increasing doses of purgatives due to annular growth at rectosigmoid junction. Always a sense of incomplete evacuation and altered bowel habits.
- **Bleeding per rectum,** frank blood or mixed with stools is common. It is painless, never massive and is the earliest symptom of carcinoma rectum. Very often, it is confused for haemorrhoids.
- **Early morning spurious diarrhoea** is due to accumulation of mucus overnight in the ampulla of rectum (dilated middle portion of rectum), which causes an urgency to pass stools but results in passage of only mucus with minimal stools. There is always a sense of incomplete defaecation.

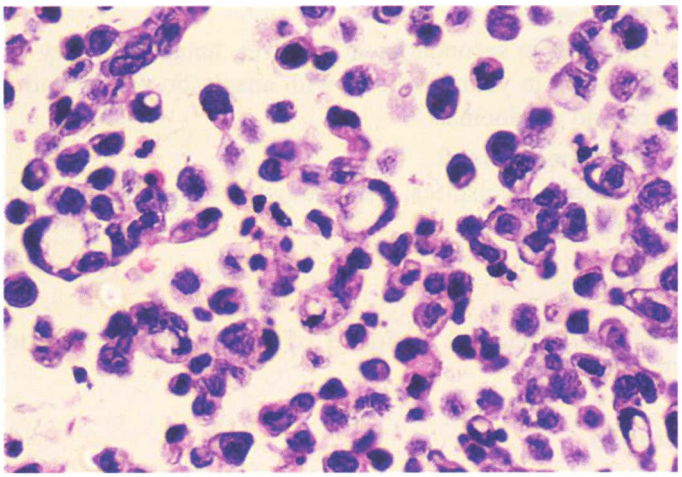


Fig. 31.12: Signet ring carcinoma colon—relatively poor prognosis (*Courtesy:* Dr Laxmi Rao, Head, Department of Pathology, KMC, Manipal)

- **Tenesmus**
  - **Painful, incomplete defaecation** associated with **bleeding** is called tenesmus.
  - This symptom is common with stricturous growths.
- **Bloody slime** (Key Box 31.3): An attempt at defaecation results in mucus mixed with blood.
- **Loss of appetite,** loss of weight due to **liver secondaries,** abdominal distension due to obstruction are late features.

ABCDEF of rectal carcinoma—symptoms

1. **A**ltered bowel habits
2. **B**leeding per rectum and bloody slime—upper rectum.
3. **C**onstipation increasing—annular carcinoma at rectosigmoid junction.
4. Incomplete **d**efaecation
5. **E**arly morning spurious diarrhoea—midrectum
6. **F**atigue, weight loss

KEY BOX 31.3

SYMPTOM	PROBABLE SITE OF LESION/EXPLANATION
• Constipation	Rectosigmoid
• Bleeding	Cauliflower-like growth
• Tenesmus	Rectosigmoid stricture
• Early morning spurious diarrhoea	Growth in the ampulla of rectum
• Bloody slime	Blood and mucus
• Sciatic-like pain	Sacral plexus infiltration
• Abdominal distension	Large bowel obstruction
• Loss of weight/ abdominal distension	Liver metastasis, ascites, etc.
• Strangury	Infiltration of the bladder base anteriorly

Clinical examination

1. **Rectal examination:** In every patient with bleeding per rectum, rectal examination has to be done. More than 90% of cases of carcinoma rectum can be diagnosed by rectal examination. Always feel for the ulcer or growth, nodularity, induration, fixity to posterior sacrum, anterior bladder base and laterally to lateral ligaments. Look for the blood stains specially in ulcerative cases. It is also possible to feel the lymph nodes in the mesorectum in cases of lower third carcinomas.
2. **Vaginal examination:** When the growth is situated in the anterior wall of the rectum, accurate assessment of the growth can be done with one finger in the rectum and the other in the vagina. Large Krukenberg tumours if present can also be felt by vaginal and rectal examinations.
3. **Evidence of metastasis:** Palpable nodular liver, para-aortic lymph nodes, ascites and supraclavicular nodes (Troisiers sign).

PEARLS OF WISDOM

Rectal cancer presenting as a fistula in ano is the equivalent to a perforated colonic cancer. It is a bad prognostic sign.

Differential diagnosis (Key Box 31.4)

1. Villous adenomas (benign) present as bleeding per rectum with mass coming out sometimes of anal canal. They have frond like appearance. They are very friable, bleeds on touch easily and they are bulky. Biopsy is a must. If it is benign, they can be removed through the rectum—submucosally.
2. Proctitis due to inflammatory bowel diseases: Both ulcerative colitis and Crohn's disease produce diarrhoea, blood in the stools and multiple ulcers. They are not indurated. Whatever it is, biopsy is a must before doing a major surgical resection. Ulcers in ulcerative colitis are typically described as pinpoint ulcers. In Crohn's they are fissure type or patchy with a cobblestone appearance.
3. **Amoebic granuloma:** It is not common nowadays. It presents with soft mass at rectosigmoid colon with or

CLINICAL NOTES

A 22-year-old girl was treated with iron tablets for anaemia due to occasional bleeding per rectum. She was treated with metronidazole because she was passing mucus along with the stools. She developed intestinal obstruction after 6 months during which time a surgeon was consulted. Rectal examination revealed a large growth, fixed all around. She died after 6 months because of advanced disease. The case illustrates the importance of rectal examination and that carcinoma of the rectum often occurs in young patients also.

KEY BOX 31.4

RECTAL ULCERS

1. Carcinoma rectum
2. Amoebic ulcers
3. Ulcerative colitis
4. HIV infections
5. Solitary rectal ulcers
6. Radiation proctitis

Fig. 31.13: Solitary rectal ulcers

without obstruction. An ulcer over the surface will mimic carcinoma. Biopsy is mandatory because amoebomas are completely curable with antiamoebic treatment.

4. **Tuberculous proctitis:** Usually patients have pulmonary tuberculosis. Submucosal abscess rupture and result in ulcers with undermined edges. Hypertrophic tuberculosis with stricture can also occur. Biopsy is mandatory before resection.
5. **Endometrioma:** It presents as constipation, bleeding per rectum especially bleeding during menstruation. Typically young females between the age of 20–40 years are affected. It produces constricting lesion in the rectosigmoid junction. Mucosa is intact as seen by sigmoidoscopy. Treatment is biopsy followed by treatment of endometriosis.

6. Solitary rectal ulcer syndrome (SRUS)

- **Site:** Commonly occurs in the anterior wall of lower rectum, an area of mucosal change.
- **Mucosa:** It is erythematous, heaped up and bleeds on touch.
- It is a single, depressed ulcer.
- The cause, even though not clear is probably due to trauma by anal digitation. Today, it is believed that it is due to **internal intussusception** or **anterior wall prolapse.**
- **Clinical features are** passage of blood and mucus in stools. Mucosal prolapse may also be a feature.
- A biopsy must be done to rule out carcinoma rectum.
- **Treatment is conservative:** Avoidance of constipation and straining may treat the prolapse.

Spread of carcinoma rectum

1. Local spread

- **It takes eighteen months** for a growth to encircle the lumen of rectum, as in annular strictures at the rectosigmoid junction.
- Then, it involves muscle coat and spreads into extrarectal tissues.
- **Anteriorly,** it involves **prostate, seminal vesicles and bladder** base in males, vagina and uterus in females.
- **Posteriorly, sacral plexus** gets involved in late cases and causes sciatica-like pain.

2. Lymphatic spread—chief nodes are para-aortic nodes.

3. Blood spread

- It results in secondaries in the liver, lungs, etc. They are common in young patients with anaplastic variety and in colloid carcinoma.

4. Peritoneal spread

- It results in ascites, carcinomatous nodules over the peritoneum, etc.

STAGING

I. Modified Dukes' staging of carcinoma of rectum (Fig. 31.14)

Stages

A Growth confined to the rectal wall
B Growth involving perirectal pad of fat and tissues. No nodes are involved.
  B1: Invading muscularis mucosa
  B2: Invading to or through serosa
C Nodes are involved
  C_1: Local lymph nodes—pararectal
  C_2: Distal lymph nodes—along the course of blood vessels
D Distant spread—liver, lungs, etc.

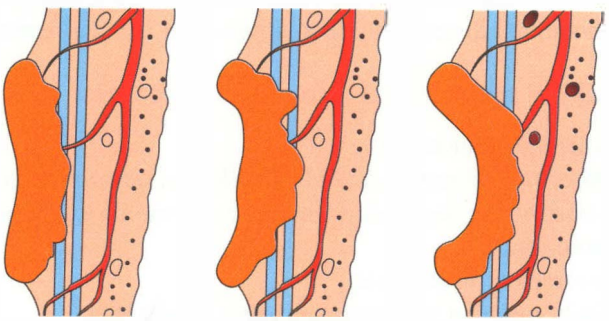


Fig. 31.14: The three cardinal stages of progression of the neoplasm

II. Astler-Coller's modification of Dukes' system

Stages

A Limited to mucosa—no nodes
B1 Extension into muscularis propria—no nodes
B2 Extension into entire bowel wall—no nodes
B3 Extension into adjacent organs—no nodes
C1 Extension into muscularis propria—positive nodes
C2 B2 + Lymph nodes
C3 B3 + Lymph nodes
D Distant metastasis

Prognosis as per Dukes' staging

- Dukes' A: 5-year survival is 90 to 100%
- Dukes' B: 5-year survival is 50 to 80%
- Dukes' C: 5-year survival is less than 50%

TNM STAGING

T represents the extent of local spread and there are four grades

T1 tumour invasion through the muscularis mucosae, but not into the muscularis propria

T2 tumour invasion into, but not through the muscularis propria

T3 tumour invasion through the muscularis propria, but not through the serosa (on surfaces covered by peritoneum) or mesorectal fascia

T4 tumour invasion through the serosa or mesorectal fascia

N describes nodal involvement

- N0 No lymph node involvement
- N1 Between one and three involved lymph nodes
- N2 Four or more involved lymph nodes

M indicates the presence of distant metastases

- M0 No distant metastases
- M1 Distant metastases

The prefix 'p' indicates that the staging is based on histopathological analysis and 'y' that it is the stage after neoadjuvant treatment, which may have resulted in downstaging

III. TNM staging

Key points in pathology, staging and spread

- They are adenocarcinomas—well differentiated, moderately differentiated and poorly differentiated.
- Signet ring carcinoma and colloid carcinoma carry poor prognosis
- Spread occurs by–local, lymphatic, haematogenous and transperitoneal routes
- Posterior sacral infiltration and anterior bladder base infiltration – surgery can be very difficult and dangerous (uncontrollable bleeding from sacral plexus of veins and prostatic plexus of veins in males. Hence preoperative chemoradiotherapy followed by surgery is done
- Involvement of mesorectum carry poor prognosis hence the **circumferential resected margin** is important (*see* page 780)
- Among the staging TNM is internationally accepted system.

Investigations (Table 31.2)

1. **Proctoscopy:** It should be done in all cases of bleeding per rectum. It is done as an out patient procedure. The left lateral position with buttocks elevated on a small pillow is the ideal position for proctoscopy. However, knee-elbow position can also be used. The growth appears as an ulcer with everted edges. A biopsy is taken to prove the diagnosis. The histological grading of the tumour is as follows:
   - A. ***Well-differentiated carcinoma: Low-grade variety (10–15%).***
   - B. ***Moderately differentiated carcinoma: The most common variety (65%).***
   - C. ***Undifferentiated carcinoma: The most aggressive variety (20–25%).***
2. **Sigmoidoscopy:** To take a biopsy from rectosigmoid growths, sigmoidoscopy is essential.
3. **Barium enema:** It is indicated when proctoscopy and sigmoidoscopy fail to give a diagnosis due to spasm of the colon. When carcinoma arises in multiple polyposis coli or ulcerative colitis, barium enema is done to rule out synchronous malignancies.
4. **Colonoscopy:** If synchronous carcinoma exists (8 to 10%) biopsy can be taken to prove the diagnosis.
5. **CEA:** Increased levels of carcinoembryonic antigen indicates metastasis.
6. **Ultrasound:** Some cases of carcinoma rectum present with metastasis such as secondaries in the liver, ascites with para-aortic nodes, etc (colloid carcinoma).
7. **Endorectal ultrasonography (EUS)** (Key Box 31.5)

   Endoscopic ultrasound staging of rectal tumors

UT1	Invasion confined to the mucosa and submucosa
UT2	Penetration of the muscularis propria but not through to the mesorectal fat
UT3	Invasion into the perirectal fat
UT4	Invasion into the adjacent organ
UN0	No enlargement of lymph nodes
UN1	Perirectal lymph nodes enlarged

8. **Computed tomography (CT) scan/MRI**
   - It helps to detect the lesion, detect metastasis in liver.
   - To know the extension of the tumour
   - To know the fixation to adjacent structures (ureter, uterus, bladder base, etc. hydronephrosis).
   - Importantly, to know nodal status

Table 31.2 Importance of each investigation and how it alters the treatment plan in a case of biopsy-proven carcinoma rectum. APR—abdomino-perineal resection, HAR, LAR—high and low anterior resection

	Plan	Investigation finding	Changed plan
1. Carcinoma lower rectum	APR	CT—metastasis in liver	Palliative colostomy/chemoradiation
2. Carcinoma upper rectum	HAR	Colonoscopy growth in transverse colon	Subtotal/total colectomy
3. Carcinoma lower rectum	APR	MRI/endosono-extensive T4 lesion	First chemoradiation followed by APR or LAR
4. Carcinoma rectum	LAR	CT scan—hydronephrosis	Cystoscopy—ureteric stenting—LAR
5. Carcinoma lower rectum	APR	PET scan—bone metastasis present	No APR

KEY BOX 31.5

EUS

- It is also called **T**rans **R**ectal **U**ltra **S**onography (TRUS)
- To know the level of penetration
- Detect perirectal lymph node enlargement
- Invasion of adjacent structures—levator ani, bony pelvis, etc.
- It is superior in T-staging of rectal cancers.

9. MRI: Both MRI and EUS are good for assessment for T-staging, MRI has following advantages

- It is better for T–3 and T–4 stage
- High resolution MRI is better for assessment of circumferential resected margin (CRM).
- It is also good for nodal staging

TREATMENT PRINCIPLES

1. Aim is to have a **curative resection**
2. **Palliative resection** is worth doing even in the presence of metastasis, when there is obstruction.
3. Even though surgical treatment is the main modality, **radiotherapy and chemotherapy** are beneficial.
4. **At surgery, ligation of vascular pedicle is done first to prevent tumour embolisation.**
5. **Ligation of bowel,** proximal and distal to the tumour helps to prevent transluminal dissemination.
6. 40% ethanol is used as tumouricidal agent to prevent suture line recurrence. Solutions such as dilute povidone iodine have been used to irrigate rectal bed after APR to prevent recurrence but without much success.
7. **Distal surgical margin should be about 2.5 cm to 3 cm.** Proximal margin—minimum 5 cm.
8. An attempt should be made to perform a **Total Mesorectal Excision (TME)**—which improves quality of life (*see* Ten commandments and Key Box 31.6).
9. **Colonic pouch:** The splenic flexure is mobilised first. A 6 cm limb of sigmoid or descending colon is folded and a pouch is created. A colotomy is made at the apex of the pouch. Linear cutter is used to staple the pouch on itself to create a common lumen. A second fire of the stapler may be necessary. This pouch now acts like a neorectum.
   - A double-stapled anastomosis as described or a hand-sewn anastomosis is then performed. A diverting loop ileostomy is used routinely for these ultra low anastomoses.

LAPAROSCOPIC MESORECTAL EXCISION

Introduction

Laparoscopic anterior resection and total mesorectal excision are well established procedures now. It is possible because of advances in the laparoscopic instruments, high definition cameras and improved technology.

TEN COMMANDMENTS OF TOTAL MESORECTAL EXCISION

1. Should do TME in all cases of mid and lower carcinoma rectum
2. Should excise the entire mesorectum (contains fat, lymph nodes and superior rectal blood vessels)
3. Should do the dissection with electrocautery or scissors
4. Should open the posterior plane between visceral and parietal layers of endopelvic fascia—**Holy plane of Heald or avascular plane**
5. Should exert good traction and counter traction to develop the planes
6. Should excise the entire mesorectum circumferentially-minimum of 5 cm of the CRM
7. Should be inside the pelvic plexus laterally
8. Should excise Denonvilliers fascia anteriorly
9. Should excise rectosacral ligament so as to reach the pelvic floor
10. Should do proximal diversion ileostomy

KEY BOX 31.6

TOTAL MESORECTAL EXCISION—ADVANTAGES

1. Mesorectum is the perirectal fat surrounding the rectum.
2. It preserves autonomic nerves
3. Impotence, urinary incontinence and retrograde ejaculation are lesser after TME.

Advantages

- A 30° camera allows a beautiful magnified view of the entire dissecting field specially low down in the pelvis. Thus, no part of the laparoscopic procedure is blind.
- Laparoscopic surgery allows the surgeon to adopt the principle of no touch technique. At the same time with better visualisation of the rectum and mesorectum all around, it permits a good dissection.
- It is not uncommon in the open method that the specimen gets torn due to traction. It has been proved that this tearing is much less in laparoscopy.
- Recovery is very fast after laparoscopy.
- Better preservation of the pelvic autonomic nerves, low anastomotic leak and low mortality rates.
- For laparoscopic low resections, one need not mobilise splenic flexure but sigmoid can be used for anastomosis.
- Wound infection, paralytic ileus are much less after laparoscopic surgery than open surgery.

Disadvantages

- **Very low resection**—because of the limited space and the lack of proper curved instruments––open method has slight advantage.

- **Port site recurrence:** Local tissue trauma due to trocars, tumour manipulation, tumour behaviour are the factors responsible for port site recurrence. Tumour spillage, tumour cell aerosolisation due to sudden loss of pneumoperitoneum, tumour spillage during extraction and immunosuppression during pneumoperitoneum are a few factors responsible for port site recurrence. This can be minimised by using bag for extraction, minimal handling of the tumour, to avoid tearing the specimen.

TREATMENT OF CARCINOMA RECTUM

Cancers arising in the distal 15 cm of the large bowel are included under this heading. These cancers behave almost like colonic cancers. The resection is the best treatment in early stages for cure. However, anatomy of the rectum, with its retroperitoneal location, narrow pelvis in males and proximity to the urogenital organs, autonomic nerves, and anal sphincters, makes surgical access and resection relatively difficult. In advanced cases neo-adjuvant therapy—chemo-radiotherapy is given to downstage the disease and then resections are done.

The treatment of rectal cancer has changed significantly over the past 20 years with the aim of cure with multimodality treatment and preserving the sphincter. Broadly the various surgery done for carcinoma rectum are as follows.

1. **Anterior resection: Refers to removal of rectum and sigmoid colon. Indicated in cases of carcinoma rectum above peritoneal reflection:** It can be low anterior resection when colorectal anstomosis is done below the peritoneal reflection or high anterior resection where the anastomosis is above peritoneal reflection. In this procedure rectum and sigmoid colon is removed along with mesorectum which contain lymphatic channels. Sphincter function is preserved. Sigmoid colon has to be removed because in cases of inferior mesenteric artery ligation, blood supply becomes inadequate and anastomosis may leak. Colorectal–colo anal anastomosis is done with or without pouch. Stapler anastomosis is the choice for low resections. Bowel is clamped and transected just proximal to this point (Fig 31.15 to 18). When the anastomosis is very low (just above anorectal ring), a protective diversion ileostomy is done which is closed after 10 weeks. Patient will have increased frequency of stools, incontinence or soilage. Diet restrictions may help these patients. Total mesorectal excision should be the aim. It improves the survival rates, decreases incidence of local recurrence.
2. **Abdominal Perineal Resection (Figs 31.19 to 22):** It is also called as Miles-Walker's operation It is indicated when the growth is too low involving the anal sphincters, poorly differentiated cancers which are very low. The patient is put in Lloyd Davis position (supine with lithotomy). Two surgeons operate simultaneously, one from the abdomen and one from the perineum. Abdomen is opened first and the growth is mobilised from the sacrum and from the urinary bladder. Pelvic dissection is carried

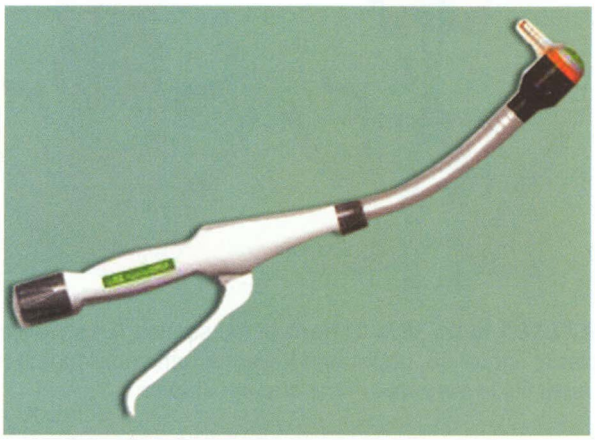


Fig. 31.16: Circular stapler used for anterior resection

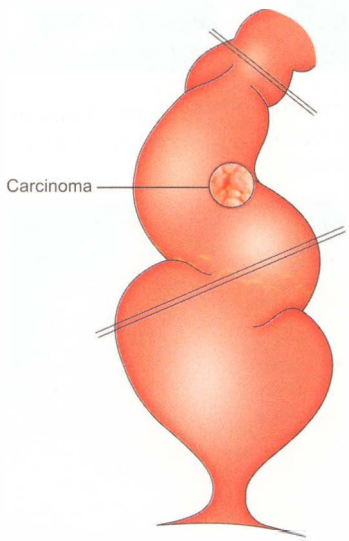


Fig. 31.15: Carcinoma upper rectum

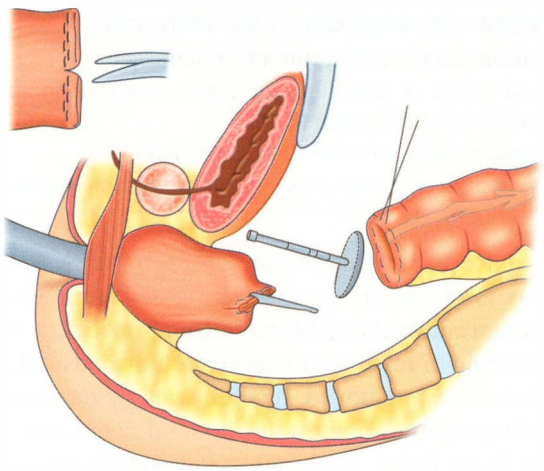
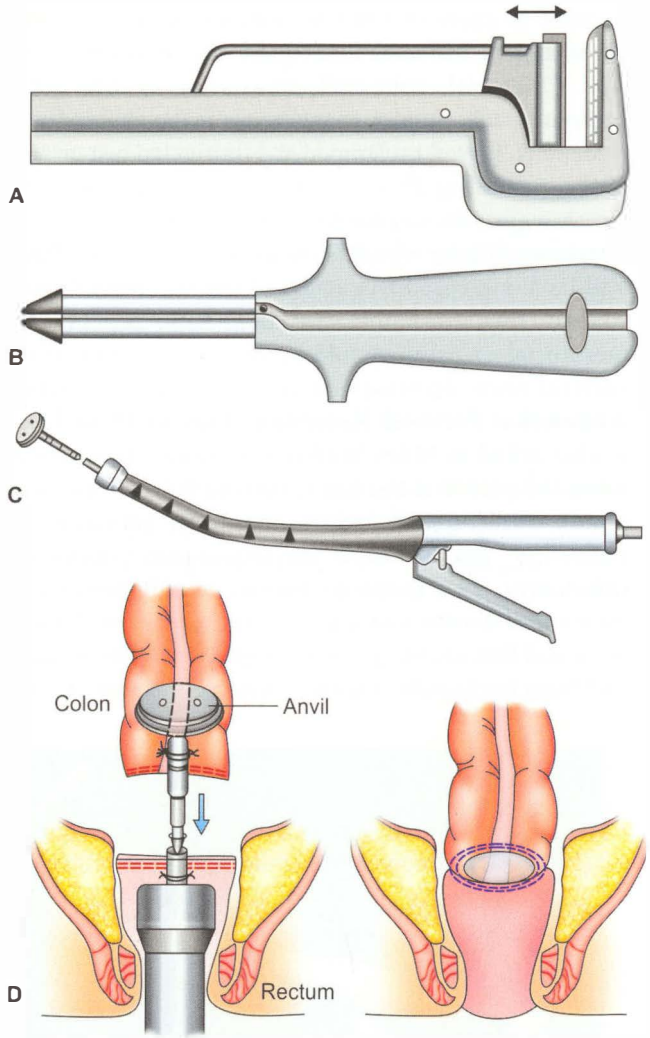


Fig. 31.17: Diagrammatic representation



Figs 31.18A to D: (A to C) Parts of the stapler, (D) shows as the stapler is tightened, upper and lower rectum come together and the anvil fits in very well in the circular stapler

KEY BOX 31.7

STRUCTURES REMOVED IN APR

- Growth with entire rectum and anal canal.
- Fascia propria with pararectal nodes.
- Two-thirds of the sigmoid colon and mesocolon with lymphatics and lymph nodes.
- Muscles and peritoneum of pelvic floor.
- Wide area of perianal skin, with part of ischiorectal fossa.

by abdominal surgeon till levator ani muscles. At this stage, anus is closed by a purse string by perineal surgeon. Rectum and anal canal is mobilized from below. The entire specimen of rectum with meso rectum and anal canal and the nodes are removed. It is followed by ***Permanent End-Colostomy*** by bringing the sigmoid colon outside in the left iliac fossa (sphincter sacrificing surgery). Thus complete excision of the rectum, anal canal, mesorectum, and lymph nodes are done through abdomino-perineal

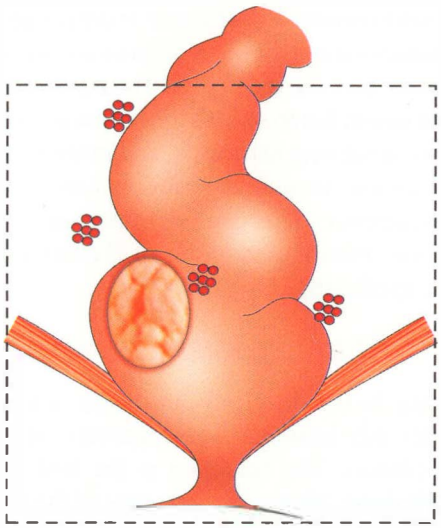


Fig. 31.19: The field of clearance in APR

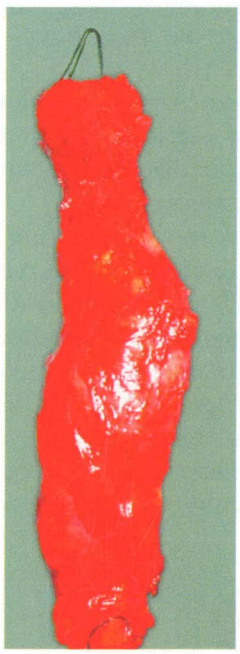


Fig. 31.20: APR specimen. Observe that the entire meso-rectum has been removed

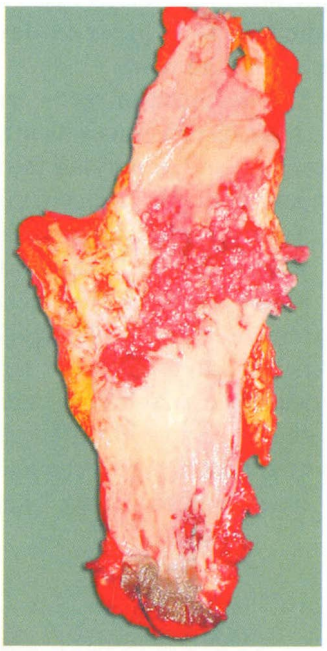


Fig. 31.21: Nodular lesion 3 cm away from the anal verge

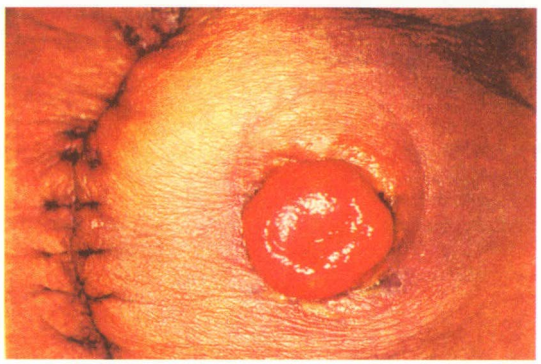


Fig. 31.22: Permanent end-colostomy following APR

VARIOUS PHOTOGRAPHS OF CARCINOMA RECTUM AT SURGERY (Figs 31.20 to 31.24)

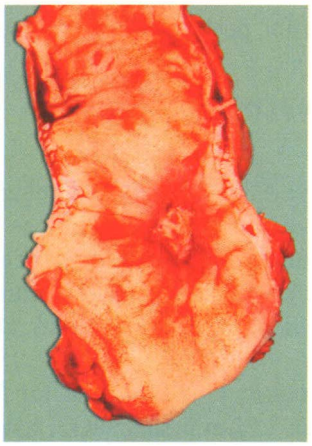


Fig. 31.23: This patient with carcinoma midrectum presented with bleeding per rectum. Low anterior resection was done. At least 3 cm margin could be achieved distally

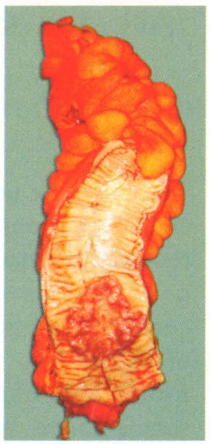


Fig. 31.24: Low anterior resection—*see* the lower margin

Fig. 31.25A: Low anterior resection is in progress. The rectum has been mobilised from urinary bladder anteriorly and sacrum posteriorly —carcinoma midrectum

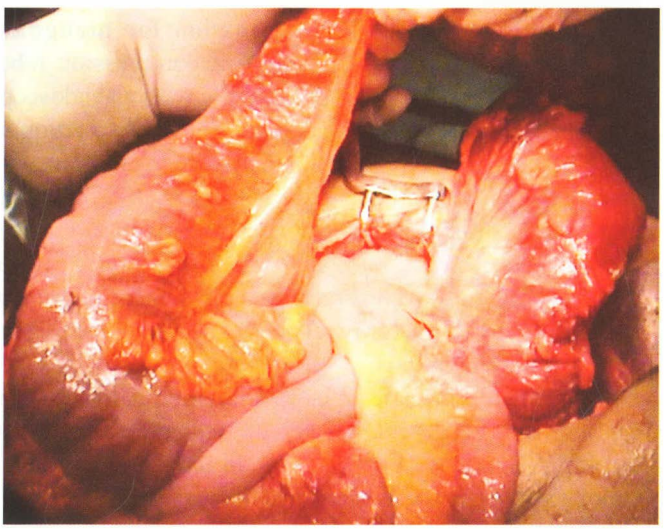


Fig. 31.25B: Proximal colon is mobilised up to the transverse colon so that it can be easily brought down without tension

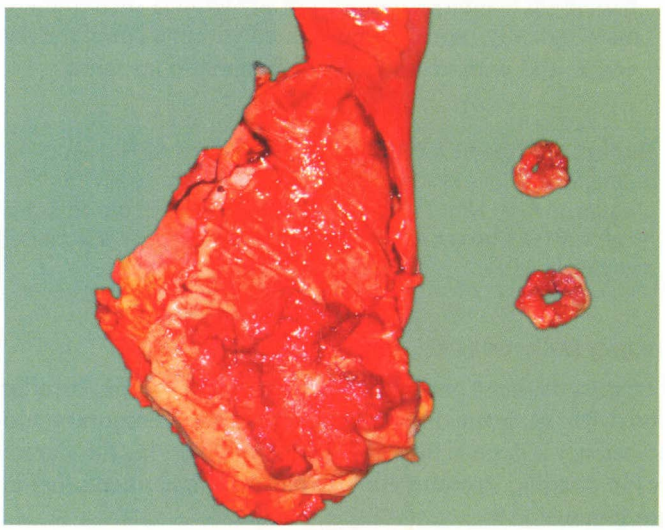
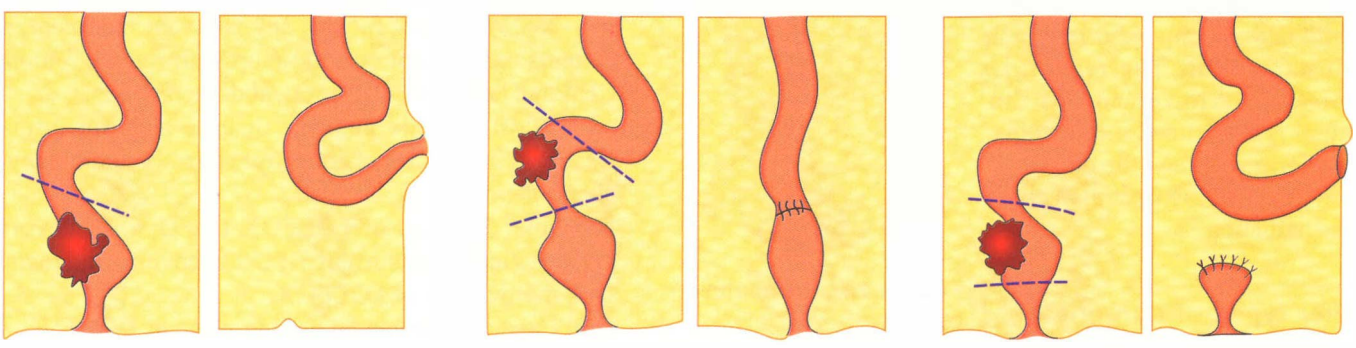


Fig. 31.26: Low anterior resection specimen—Stapler anastomosis has been done—you can see the complete doughnuts



Figs 31.27A to C: Schematic representations of resection of the rectum. (A) AP resection followed by colostomy, (B) High anterior resection followed by anastomosis, (C) Lo w anterior resection with colostomy and closure of rectal stump—Hartmann's procedure—in large bowel obstruction. The colostomy is closed and colorectal anastomosis is done after 4 weeks

KEY BOX 31.8

LOCAL EXCISION OF CARCINOMA RECTUM

- Mobile tumours less than 4 cm in diameter
- Less than 40% of rectal wall involvement
- Located within 6 cm of anal verge
- Lesion should be T1 or T2
- No vascular or lymphatic invasion
- No nodal involvement—preoperative MRI or EUS.

incision. Usually one drainage tube is inserted brought out through perineal wound.

3. **Local excision:** If the tumor is confined to the submucosa (uT1, N0), excision can be done by a transanal approach. The explanation being these cases will have less than 8%,lymphatic metastasis. In our country majority of patients do not fall in this category. It is indicated for mobile early tumors that are less than 4 cm in diameter, that involve less than 40% of the rectal wall circumference, with good histology like well or moderately differentiated histologically, and that are located within 6 cm of the anal verge. and with no vascular or lymphatic invasion.

Recent change in APR: In the conventional APR dissection is within the levator ani and levator ani is preserved. The change is APR with removal of levator ani—it is called extra levator APR. The resected specimen has a waist—it is called cylindrical APR.

Inoperable cases

Locally advanced growths present with severe pain, bleeding and with subacute intestinal obstruction. Temporary loop colostomy is done in the left iliac fossa by bringing the sigmoid colon outside. Postoperatively, radiation and chemotherapy are given.

Hartmann's operation

(Fig. 31.28)

This is indicated in old and debilitated patients who may not withstand APR. The rectum is excised, the lower end of the rectum is closed and a colostomy is performed. When the growth is slow-growing, this operation gives good palliation.

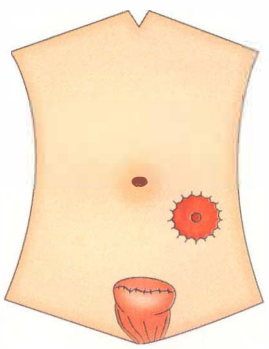


Fig. 31.28: Hartmann's operation

Other types of surgeries

- **TEM: Transanal endoscopic microsurgery (BUESS)**
- It involves wide retraction, followed by good visualisation of lesion followed by complete thickness excision and direct suturing. It can be done by transanal, trans-sacral or trans sphincteric approach.
- **Trans-sacral (Kraske's)** is for posterior rectal tumour.

ENDOSCOPIC TREATMENT OF COLORECTAL LESIONS

- **Colonoscopic polypectomy:** All adenomas and potentia adenomas should be removed. Pedunculated polyps are removed by snare. Small polyps can be removed by cold or hot forceps and hot or cold snare. Bleeding can be controlled by injection of epinephrine, electrocautery, clips or endoloops.
- **Endoscopic mucosal resection:** It is indicated for flat, sessile lesions, not more than 2.5 cm in diameter. First the lesion is elevated by injecting saline or hypertonic dextrose with epinephrine. The lesion is elevated, cut and removed or sucked and cut.
- **Endoscopic submucosal dissection:** Technically more difficult. Principle of the procedure is same. More chances of perforation.
- **Endoscopic stents for decompression for malignant obstruction:** Large bore colorectal decompression tubes are available. They can be passed with or without endoscope with a guidewire. They are used to relieve obstruction so that bowel preparation can be done and one stage treatment—resection and anastomosis can be done. Self expandable metal stents—SEMS has been used as palliation in cases of large gut obstruction. Quality of life is slightly better but no increase in survival rate of patients. Stent migration, tumour ingrowth, overgrowth, perforation and bleeding are the other complications.
- **Laser therapy:** Like oesophageal carcinoma, lower rectal carcinoma with obstruction can be treated as a palliation by Nd:YAG laser. It cannot be used in acute cases. Complications are perforation, bleeding, fistula formation, etc.

ROLE OF RADIOTHERAPY AND CHEMOTHERAPY

Postoperative Management

1. pT1-2N0M0 do not require any adjuvant treatment, such patients can be kept on follow-up with routine 3 monthly CEA and annual CECT thorax/abdomen/pelvis.
2. pT3N0M0 or node positive disease requires adjuvant treatment in the form of concurrent chemoradiotherapy and chemotherapy.
   Example: 2 cycles of FOLFOX (5-FU + Leucovorin + Oxaliplatin) → concurrent 5-FU/Leucovorin and radiation → 2 more cycles of FOLFOX.
   Oral Capecitabine can be used in place of IV 5-FU.
3. It is preferable to add Oxaliplatin in the chemotherapy regimen if nodes were positive for metastatic disease. Although in older population (> 65–70 years) it might be of less benefit.

4. Radiation portals should include the postoperative tumor bed, pre-sacral nodes, internal iliac nodes. External iliac nodes can also be included in T4 tumors.
   Radiation dose is usually 45–50 Gy given over 5 days a week for 5 consecutive weeks. Another 5–9 Gy boost to the tumor bed ONLY can be considered especially with adverse features like lymphatic emboli, close margins.

Preoperative—Neoadjuvant chemoradiation
(Key Box 31.9)

Rationale: In locally advanced cases where the surgeon feels that complete resection may not be feasible or sphincter saving will not be possible, neoadjuvant chemoradiation can be attempted thus saving the patient from having a permanent colostomy bag and also a better curative outcomes.

KEY BOX 31.9

ADVANTAGES OF PREOPERATIVE RADIOTHERAPY

- Decreased tumour seeding at surgery
- Increased radiosensitivity due to more oxygenated cells
- Conversion of APR to LAR

- Generally clinically T3-4 tumors which may or may not be node positive are eligible candidates for neoadjuvant chemotherapy.

1. A typical course of neoadjuvant therapy comprises concurrent 5-FU/Capecitabine and radiation. A dose of 45–50 Gy is used to treat the pelvis including the growth and the draining lymphatic regions followed by 5 Gy boost to the tumor itself.
2. Following neoadjuvant therapy, patient should be re-evaluated using CT/MRI for possibility of resection.
3. Surgery is usually considered after 6–8 weeks following neoadjuvant therapy as the maximal response to the treatment may take up to 2 months.
4. Further adjuvant treatment is to be given following surgery depending upon the histopathological report.

PEARLS OF WISDOM

Rectal cancers are more radiosensitive and colonic cancers are more chemosensitive.

LOCALLY RECURRENT RECTAL CANCER

- Major cause is a positive margin on pelvic side wall. This is the reason why preoperative chemoradiotherapy should precede excision of T3 and T4 lesions with TME.
- Usually develops within 18 months.
- Presents as pelvic pain, mass and rectal bleeding.
- Pelvic CT, MRI, CEA levels are the required investigations.
- Chemoradiation, surgery, local palliative treatment, pelvic exenteration (resection of rectum and bladder—Brunschwig's operation) are alternative treatments available.

COLOSTOMY

Opening of the colon to the exterior, either temporary or permanent, for the drainage of faecal matter is called colostomy.

TYPES

1. Temporary colostomy

A. In cases of acute left-sided colonic obstruction, proximal half of right transverse colon is brought out through the upper part of the right rectus abdominis muscle. Later, radical resection of the left colon is done followed by closure of the colostomy.

B. In cases of traumatic or congenital fistula affecting the left colon, temporary colostomy is indicated (Fig. 31.36). The loop of the colon which is brought outside is held in place by a glass rod. This is passed through the transverse mesocolon and held by rubber tubing. This rod is removed after 10 days.

C. Colostomy/Ileostomy is done if a distal colorectal anastomosis gives way (Figs 31.29 and 31.30).

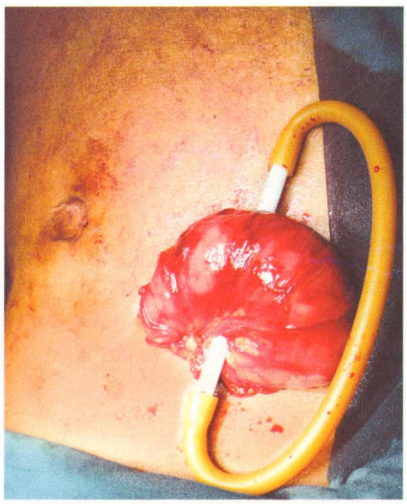


Fig. 31.29: Sigmoid colon is brought out and a glass rod connected to rubber tube is used to hold it out

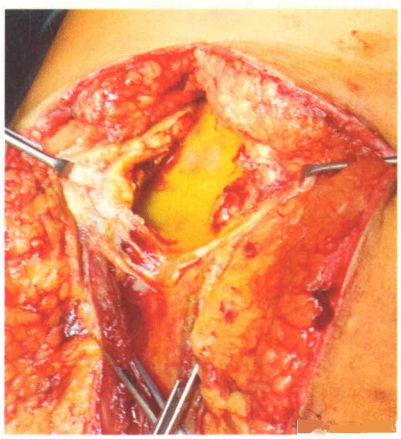


Fig. 31.30: Faeculent leak from failed anterior resection

PEARLS OF WISDOM

Transverse colostomy is bulky, contents are semiliquid and difficult to manage—Try to avoid it.

TEN COMMANDMENTS

1. Should mark the colostomy site in the standing and sitting positions
2. Should be at least 3 cm away from the bony landmark (anterior superior iliac spine)
3. Should be at least 3 cm away from the midline incision. Otherwise, colostomy will contaminate the incision site (Fig. 31.31)
4. Should close the paracolostomy space within the abdomen to prevent herniation of the small intestines
5. The colostomy should be matured after the midline incision is completely closed and sterile dressings applied
6. Should be in flush with skin surface in the left iliac fossa
7. Should excise a disc of skin for a permanent colostomy
8. The stoma should be brought out through the rectus muscle
9. Should avoid bringing stoma outside through scar tissue
10. Should examine the end of the colostomy for vascularity (Fig. 31.32)

2. Permanent colostomy

It is indicated after abdominoperineal resection where the end of sigmoid colon is brought outside in the left iliac fossa as permanent colostomy (Figs 31.31 and 31.32, Key Box 31.10). The colostomy site should be 3 cm away from anterior superior iliac spine so that colostomy bag can be fitted properly (Figs 31.33 and 31.34).

3. Double-barreled colostomy

In this the adjoining walls of the intestine are crushed. Both ends of the loop are defunctioned. This type of colostomy is not frequently done now. It was done earlier for sigmoid volvulus, resection of colonic stricture, etc.

Indications for colostomy

- **Congenital:** In Hirschsprung's disease and anorectal anomalies, temporary colostomy is done first.
- **Carcinoma:** Following APR, permanent end-sigmoid colostomy is done.
- **Colonic fistulae:** Fistulae due to diverticulitis, Crohn's disease or due to tuberculosis.
- **Colonic injuries:** Trauma due to stab injuries or due to operative injuries following nephrectomy, pelvic operations, PCNL (percutaneous nephrolithotomy).

Advantages of colostomy

Distal bowel takes complete rest, regains normal size and bacterial colonisation is reduced. It becomes empty and sterile so that chances of leakage at a later operation is reduced.

Fig. 31.31: End colostomy away from the main incision

Fig. 31.32: Inspect for vascularity of colostomy following APR

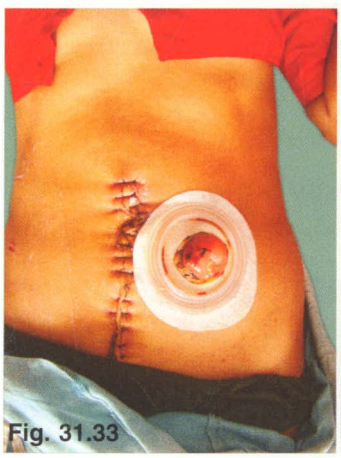


Fig. 31.33

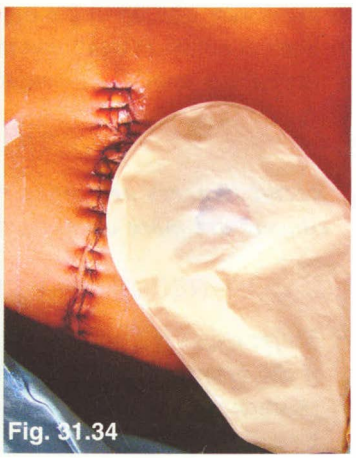
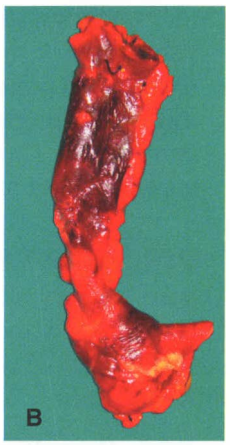


Fig. 31.34

Fig. 31.33: Disposable colostomy bags are available with or without flange. Depending upon the size of the stoma, it can be cut open so as to fit into the colostomy, **Fig. 31.34:** A colostomy bag has been applied. It can be reused

Fig. 31.35: Paracolostomy hernia: Herniation of bowel from the side of colostomy



Figs 31.36A and B: Colostomy gangrene. It is mandatory to inspect the end of the colsotomy everyday in the postoperative period to check for vascularity. If it is gangrenous and if it is retracted it is better to open the patient again and refashion colostomy

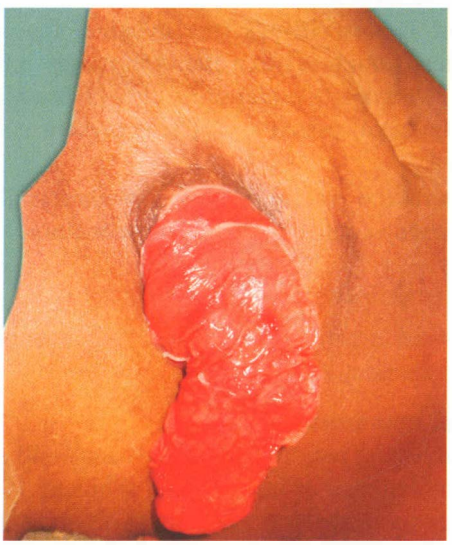


Fig. 31.37: Colostomy prolapse because sutures would have given way where it was anchored to aponeurotic layer. The patient also had violent postoperative cough

Fig. 31.38: Burst abdomen due to contamination of the wound from colostomy—proper care should be taken to isolate laparotomy site and colostomy till complete healing of laparotomy wound takes place

Complications of colostomy (Figs 31.35 to 31.38)

- Bleeding, necrosis, retraction, prolapse, parastomal hernia and colostomy diarrhoea are some complications.
- Colostomy obstruction, gangrene.

PEARLS OF WISDOM

Colostomy should be done by an experienced surgeon as patient has to live with the colostomy life long.

KEY BOX 31.10

PERMANENT COLOSTOMY—LEFT ILIAC FOSSA

- Avoid bony prominences
- Avoid belt lines
- Avoid marking in lying down position.
- Avoid scars
- Avoid bringing out stoma lateral to rectus.

PROLAPSE RECTUM

Protrusion of the mucous membrane or the entire rectum outside the anal verge. This condition is common in children and elderly patients.

Types

Prolapse can be of two types: Partial prolapse and complete prolapse.

Partial prolapse

- In this variety, the protrusion is between 1.25 and 3.75 cm outside the anal verge (Fig. 31.39).
- It is usually a mucosal prolapse.

Fig. 31.39: Partial prolapse

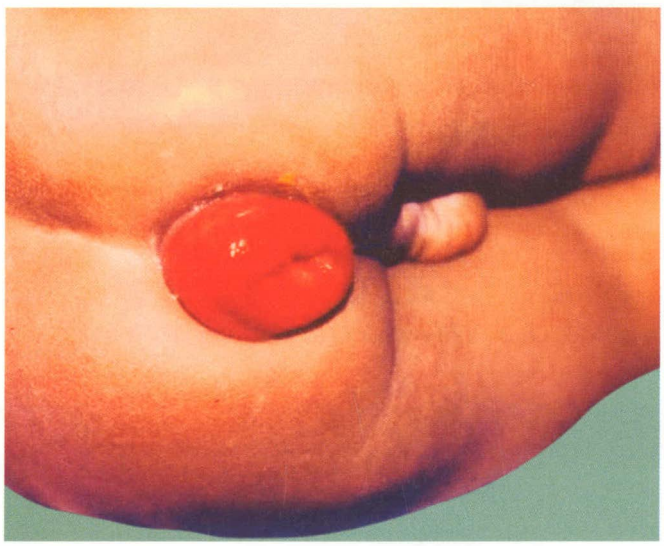


Fig. 31.40: Rectal prolapse in a child

Causes

1. In infants, it is due to **undeveloped sacral curve** and in children it can be secondary to **habitual constipation.**
2. It can follow an attack of **whooping cough or excessive straining** (Fig. 31.40).
3. It can follow an attack of diarrhoea resulting in **loss of fat** in the ischiorectal fossae, which supports the rectum.
4. In adults it is common in females mostly due to **Torn perineum** caused by obstetric trauma.

Treatment

I. **Digital reposition:** In infants, partial prolapse is temporary. The mother is advised to push the prolapse inside after lubricating with lignocaine jelly.
2. **Injection of ethanolamine oleate** into the submucosa of the rectum. It causes aseptic fibrosis. Thus, mucosa gets tethered to the other layers.
3. Partial prolapse can be **excised**, after applying **Goodsall's ligature.**

COMPLETE/TOTAL PROLAPSE

- Full thickness prolapse is also called procidentia.
- It is defined as protrusion of the rectum for more than 3.7 cm outside the anal verge. Very often, it is the entire rectum which protrudes out on straining, sometimes along with the peritoneal sac.
- Often, it is associated with prolapse uterus.

The pelvic floor—surgical anatomy (Fig. 31.41)

- It is composed of the **two levator ani** and a **puborectalis** muscle.
- Levator ani originate from pelvic side walls and sacrospinous ligament. It **suspends** the rectum in a muscular sling which ends when puborectalis angulates rectum.
- Puborectalis muscle takes origin from posterior aspect of pubis, forms a sling around rectum and return to posterior pubis.
- **Contracted puborectalis is responsible for normal acute anorectal angle and it is critical for maintaining continence.** Thus during coughing, sneezing, anorectal angle becomes more acute, increasing continence.

Supports of the rectum and surgical importance

Various supports of the rectum keeps the rectum in place. Failure of one or more of these factors may precipitate rectal prolapse (Fig. 31.42). They have been enumerated in the following lines.

1. **Pelvic floor**: Weakness of pelvic floor can be due to birth injuries or due to defective collagen maturation.
2. **Lateral ligaments**: These ligaments are due to condensation of pelvic fascia on each side of the rectum. Excessive mobility of these ligaments may be the contributing factor for prolapse rectum.
3. **Fascia of Denonvilliers** (rectovesical fascia): **Deep rectovesical pouch** is often found in prolapse rectum. In all cases of complete prolapse rectum, please look for the deep rectovesical pouch and if it is present it should be obliterated.
4. **Fat** supports the rectum. Hence, any chronic illness and loss of fat may contribute for prolapse rectum.

Anorectal physiology and investigation

These are useful in patients who have complaints of prolapse rectum, constipation, incontinence.

1. Anorectal manometry

- Normal resting pressure in the anal canal—40–80 mmHg (it is the function of internal anal sphincter.)
- **Squeeze pressure**: It is maximum voluntary contraction pressure minus resting pressure. It is 40–80 mm above resting pressure. It reflects function of external anal sphincter.

A FEW PHOTOGRAPHS OF PROLAPSE RECTUM

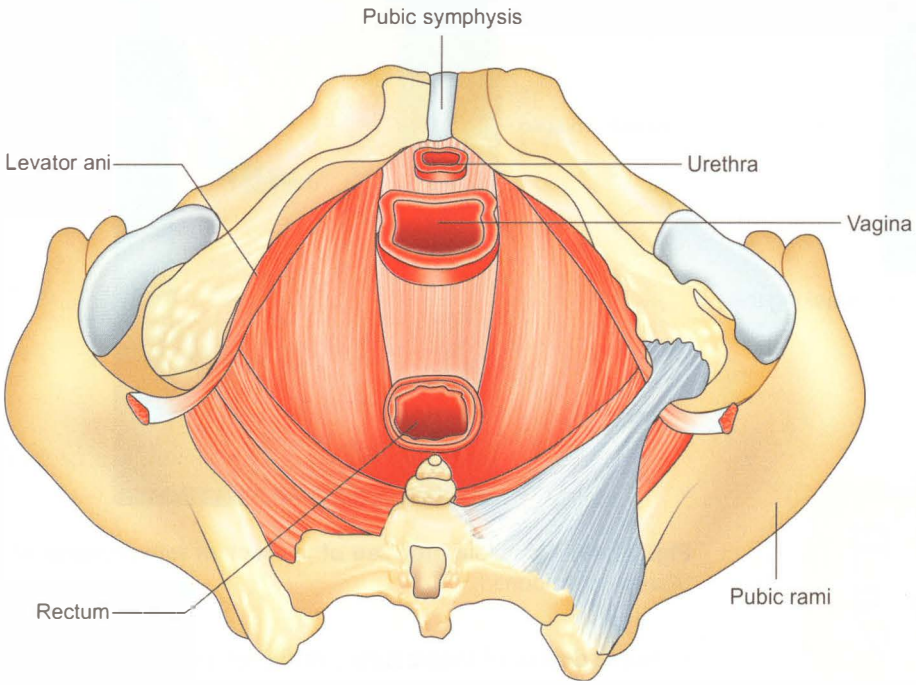


Fig. 31.41: Pelvic floor anatomy—weakness of the pelvic floor is an important cause of prolapse rectum

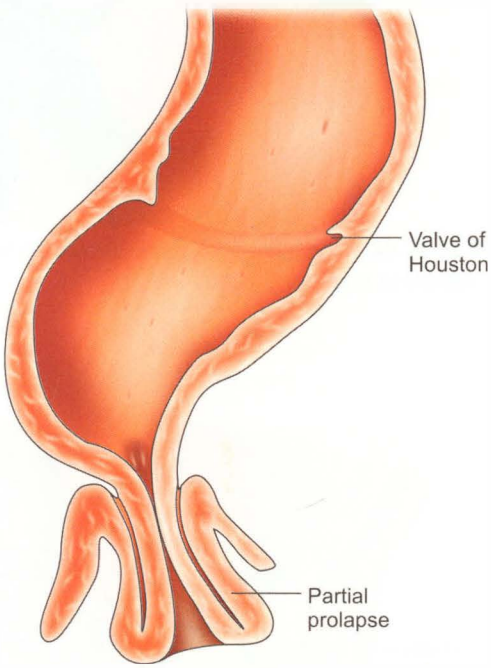


Fig. 31.42: Prolapsed rectum—diagrammatic representation, sometimes confused for prolapsed haemorrhoids

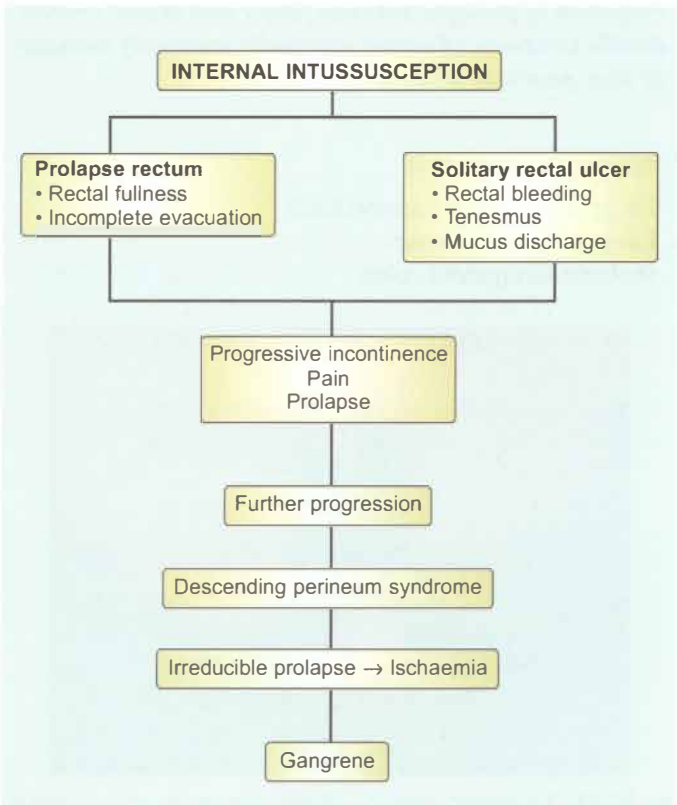


Fig. 31.43: Pathogenesis of prolapse rectum

2. **Function of pudendal nerve and puborectalis nerve:** These are nerve conduction studies which can reflect injuries to the nerve.

Causes (Fig. 31.43)

1. **Common in elderly women who are multipara.** Probably, it is due to repeated **birth injuries** to the perineum causing damage to the nerve fibres. As age advances, muscles become weak. This, together with fatty degeneration of the muscle, results in prolapse rectum.
2. **Excessive straining** causes weakness of the supports of the rectum.
3. **Defective collagen maturation** results in failure of rectal support by levators and pelvic fascia.
4. Presence of **deep rectovesical pouch** and excessive mobility of the rectum (mesorectum) predisposes to prolapse of the rectum.
5. Many people believe that prolapse of the rectum starts as an **intussusception** in the first stage, initiated by certain factors such as diarrhoea, constipation and disorder of the pelvic floor. The process starts with anterior wall of rectum, where supporting tissues are weakest (Fig. 31.45).

Fig. 31.44: Complete prolapse—should be tested by asking the patient to squat and strain

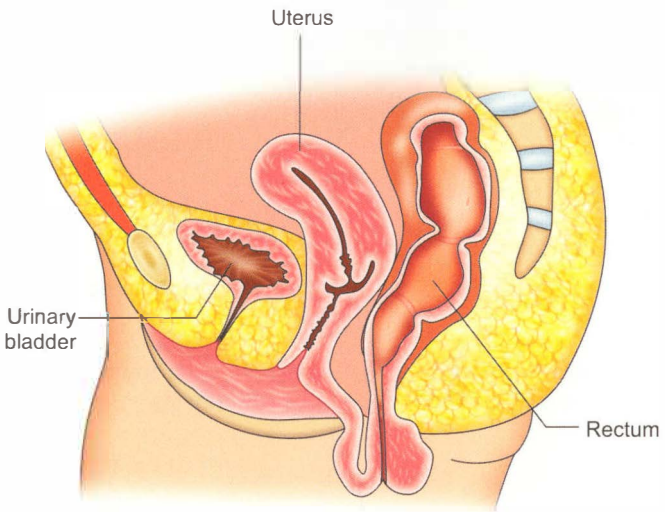


Fig. 31.45: Prolapse of the rectum with uterus: Procidentia—not an uncommon presentation specially in multipara women

KEY BOX 31.11

OBSTETRIC TRAUMA

- Multiple vaginal deliveries—cause pudendal nerve stretch.
- Prolonged labour—disrupts sphincter and stretching of pudendal nerve.
- 3rd degree perineal tears—weaken the internal sphincter and pelvic floor.

Clinical features

- Female-male ratio is 6 : 1.
- Constipation is an important feature of rectal prolapse.
- Excessive mucus discharge causing irritation to the perianal skin. Tenesmus is also common.
- On asking the patient to strain at stool[1], the rectum descends down, which clinches the diagnosis (Fig. 31.46).

[1]In fact, the surgeon should see the prolapse of the rectum outside when patient strains to make a clinical diagnosis.

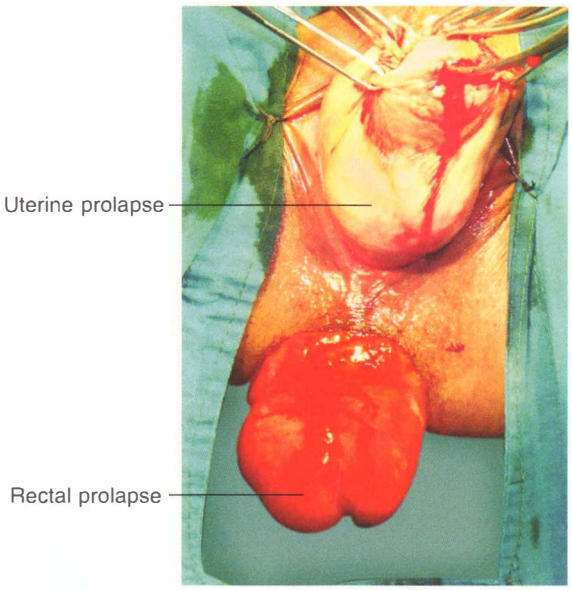


Fig. 31.46: Complete prolapse of the rectum with prolapse of uterus—procidentia

- Some degree of incontinence of faeces and flatus is always present. It gives rise to urgency and perianal soiling.
- Rectal examination—lax anal sphincter and wide gaping on straining.
- Procidentia (Fig. 31.47)

PEARLS OF WISDOM

Palpation of prolapse between finger and thumb reveals double thickness of tissue, especially anteriorly because of deep pouch of Douglas.

Differential diagnosis

- Large third degree haemorrhoids
- Large polypoid tumour
- Prolapse of sigmoid colon

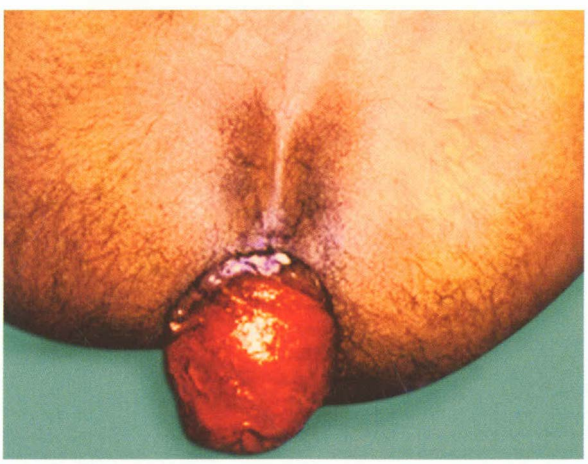


Fig. 31.47: Prolapsed rectum—slowly change in colour—would have gone for gangrene

Complications

- Proctitis, ulceration and rarely bleeding
- Gangrene of the rectum (Fig. 31.48)

Treatment: Surgical procedures—aim

1. Safe procedure to correct with minimal morbidity and without mortality. They are classified as **perineal procedures and abdominal procedures**.
2. To cure or to improve **incontinence.**

I. PERINEAL PROCEDURES

1. **Delorme's procedure** (reefing the rectal mucosa): In this, the prolapse is completely everted, mucosa is stripped and muscle coat is plicated. Mucosal continuity is maintained by suturing anal canal mucosa below to the rectal mucosa above. This is an easy operation to do in elderly patients. However, relapse rates are high and it does not correct the defect.
2. **Altemeier's procedure:** In this operation, full thickness of the prolapsed rectum with part of sigmoid is ***excised*** followed by ***anastomosis of part of the sigmoid*** to the ***anal canal*** from below. To improve continence, plication of levator ani and puborectalis muscle is done. ***Urgency and incontinence*** are the features because of removal of rectum.
3. **Thiersch wiring:** In this operation, a steel wire or a thick silk suture is applied all around the anus after reducing the prolapse. The knot is tightened around a finger. Patients with poor surgical compliance benefit from this operation. However, breakdown of the wire, perianal sepsis and anal stenosis are the complications.

II. ABDOMINAL PROCEDURES

1. **Wells operation:** A laparotomy is done, rectum is pulled upwards and is sutured to the sacrum posteriorly with the help of a polyvinyl alcohol sponge kept behind the rectum. The sponge is sutured posteriorly and laterally to the walls of the rectum. Dense fibrotic reaction occurs resulting in fixation of the rectum to the sponge.
2. **Ripstein sling operation:** After a laparotomy, the rectosigmoid junction is sutured to the sacrum by using **Teflon sling**, below the sacral promontory. One complication of this operation is constipation due to rectosigmoid angulation. Hence, sigmoidectomy has been suggested along with this operation.
3. **Mesh rectopexy:** Instead of polyvinyl sponge, a marlex mesh can be kept behind the rectum. This is sutured behind, to the sacrum and then to the posterior and lateral surfaces of rectum. **Laparoscopic method of fixing the mesh has become popular. This is the procedure of choice today.** Constipation is one of the complications of mesh rectopexy. Hence, some resect sigmoid with this procedure **(Goldberg operation)** (Fig. 31.49).
4. **Lahaut's operation:** Anchoring rectosigmoid to rectus sheath (extraperitonisation).

Key Box 31.12 shows summary of surgeries for prolapse rectum.

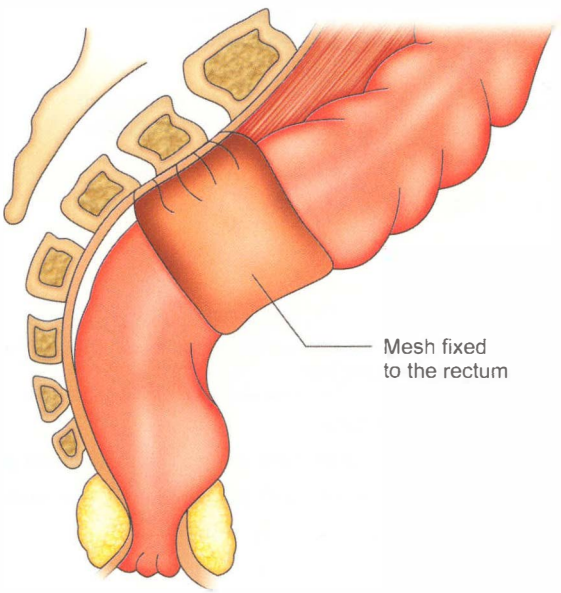


Fig. 31.48: Fixing the mesh to the rectum—mesh is sutured to the sacrum posteriorly and sutured to the sides of the rectum

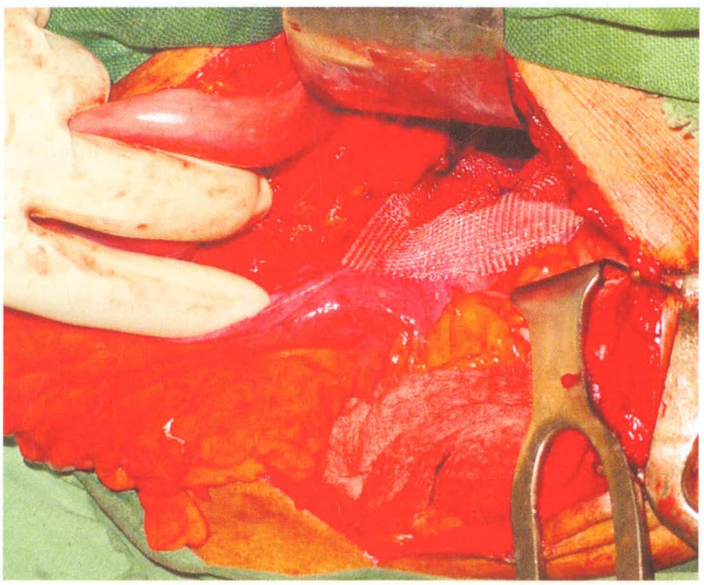


Fig. 31.49: Mesh fixation for total prolapse of the rectum—most common surgery—open or laparoscopic method

KEY BOX 31.12

SUMMARY OF SURGERIES FOR PROLAPSE RECTUM

- **Mesh rectopexy** corrects/prevents prolapse but does not correct chronic constipation.
- Laparoscopic mesh rectopexy has become **gold standard**—fast recovery, less pain, short hospital stay.
- **Mesh rectopexy with resection** is ideal for patients with constipation or patients with a redundant sigmoid colon.
- High operative risk patients—**Thiersch wiring**—anal encirclement.
- **Altemeier** procedure done for perineum is an alternative in patients with **incontinence**. Here, perineal proctectomy and posterior sphincter enhancement is done.

SURGICAL ANATOMY OF ANAL CANAL

It is 3 cm long, starts as the continuation of rectum, passes through pelvic diaphragm and ends at the anal verge (skin).

Internal sphincter (Fig. 31.50)

- It is the continuation of circular muscle fibres of rectum and ends 0.5 cm below pectinate line.
- It is involuntary and 2.5 cm long.
- Internal sphincter with fibres of external sphincter and puborectalis which maintain the anorectal angle, form the **anorectal bundle, and maintains continence.**
- Its fibres are transversely placed. Motor fibres come from presacral plexus.

External sphincter

- It is formed by **striated muscle fibres** intermingled with longitudinal muscle fibres of the rectum which get attached to the skin of perianal region.
- It has superficial, deep and cutaneous portions.
- Levator and puborectalis have an attachment with internal sphincter.
- Nerve supply (motor) comes from inferior haemorrhoida branch of internal pudendal nerve and perianal branch o the 4th sacral nerve (motor to levator ani also).
- It is voluntary and gives temporary continence.

Development

- Anal canal is developed from fusion of **post-allantoic gut** with **proctodeum**.
- The junction of these is the dentate line or pectinate line. **Anal valves of Ball** are remnants of proctodeal membrane.
- At the level of dentate line, the mucosa is folded in the form of longitudinal columns—***columns of Morgagni.***
- In between the columns of Morgagni, 4–8 anal glands open into the small anal sinuses.

Comparison of anal canal above and below the dentate line is given in Table 31.3.

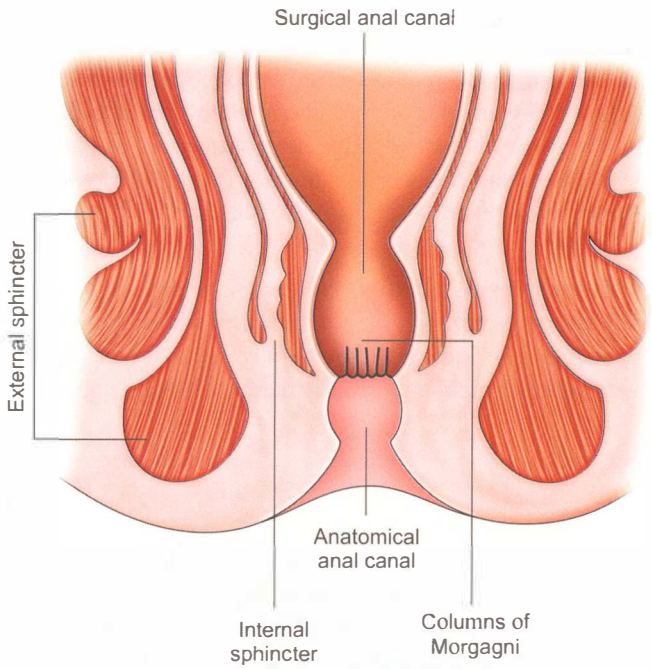


Fig. 31.50: Anatomy of the anal canal

Table 31.3 Comparison of anal canal above and below the dentate line

	Above the dentate line	Below the dentate line
1. Nomenclature	Surgical anal canal	Anatomical anal canal
2. Epithelium	Cuboidal epithelium	Skin—squamous epithelium, without hair and sweat glands
3. Nerve supply	Parasympathetic. Hence, painless	Spinal nerves, inferior haemorrhoidal nerve, very painful
4. Venous drainage	Portal system	Systemic veins (external iliac vein)
5. Colour	Pink	Skin colour
6. Development	Post-allantoic gut	Proctodeum
7 Lymphatic drainage	Para-aortic nodes	Superficial and deep inguinal nodes

HAEMORRHOIDS (PILES)

Definition

Dilated plexus of superior haemorrhoidal veins, in relation to anal canal.

Classification—aetiological

I. Primary/Idiopathic haemorrhoids

1. **Standing posture:** It has been told nicely that varicosity is the penalty for verticality against gravity. It is also true for haemorrhoids. It is true that animals do not develop haemorrhoids.
   Thus man's upright posture and absence of valve in the portal system with other factors precipitate development of haemorrhoids
2. **Haemorrhoidal veins** and their branches are the thin veins which pass through submucosa of the rectum. They get compressed due to contractions caused by rectal musculature (the sphincters) during the act of defaecation.
3. **Genetic/ familial factors:** Absence of valves, or congenital weakness of the vessel wall are few other factors contributing for the haemorrhoids
4. **Diet:** A diet deficient in fibres which prolongs the gut transit results in constipation and small hard pellet like stools. The hard stools compress veins and result in haemorrhoids.

II. Secondary haemorrhoids

Causes

1. **Carcinoma of rectum**, by blocking the veins, can produce back pressure and can manifest as piles.
2. **Portal hypertension**—uncommon cause of rectal varices.
3. **Pregnancy**, due to compression on superior rectal veins or due to progesterone which relaxes smooth muscle in the wall of the veins, can cause haemorrhoids.

   Current view: Latest theory is that haemorrhoids occur due to caudal displacement of anal cushions. It is due to recurrent trauma, shearing forces, loss of elasticity. Thus normally the cushions retract after defaecation.

Location

Classically situated in the 3, 7, 11 o'clock positions (Fig. 31.51) (left lateral, right posterior and right anterior respectively).

- Superior haemorrhoidal artery (vein) gives 2 branches on right side and 1 branch on left side. Hence, piles are two on right side and one on left side.

Clinical features (Table 31.4)

- Painless bleeding—fresh bleeding occurs after defaecation **Splash in the pan.** This causes chronic anaemia. Haemorrhoids which bleed are called Grade I haemorrhoids.
- The capillaries of the lamina propria are only protected by a single layer of epithelial cells. Hence, minor trauma precipitates bleeding.

Table 31.4 Grades of haemorrhoids

Grades	features	Symptoms
I.	Never prolapse	Bleeding per rectum
II.	Prolapse on defaecation Spontaneous reduction	Something coming down and going back
III.	Prolapse on defaecation requires manual reduction	Something coming down, bleeding, mucus discharge, pruritus
IV.	Permanent prolapse	Acute pain, throbbing discomfort

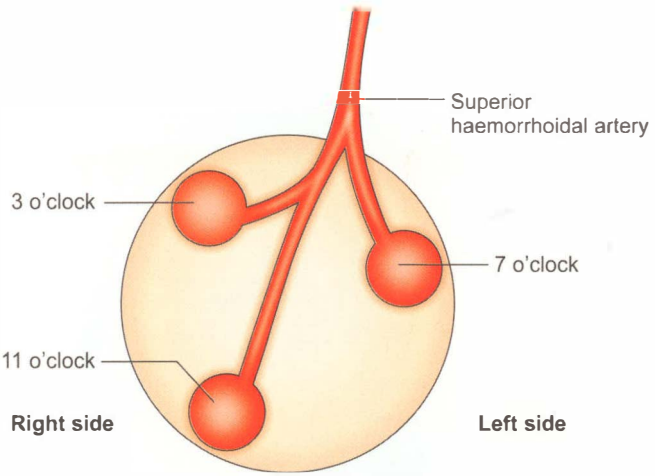


Fig. 31.51: Classical location of pile masses

- As the straining increases, the haemorrhoids partly prolapse outside. After defaecation, it returns back (Grade II) or can be digitally replaced (Grade III haemorrhoids).
- Permanently prolapsed pile outside (Grade IV haemorrhoids). The patient complains of pain or discomfort.
- Most of the patients complain of constipation.
- Discharge of mucus and soiling of perianal skin—pruritus by prolapse of haemorrhoidal cushions and mucosa (Figs 31.52 to 31.54).

III. Depending upon the location of haemorrhoids

1. **Internal haemorrhoids**—above the dentate line, covered with mucous membrane.
2. **External haemorrhoids**—at anal verge, covered with skin (Fig. 31.54).
3. **Interno-external**—both varieties together.

Investigations

- Per rectal examination is done mainly to rule out carcinoma rectum or other causes of bleeding per rectum. Haemorrhoids cannot be felt by rectal examination unless they are thrombosed or fibrosed.
- Proctoscopy: As the obturator is removed, piles prolapse into the lumen of proctoscope as cherry red masses.

Fig. 31.52: Grade III

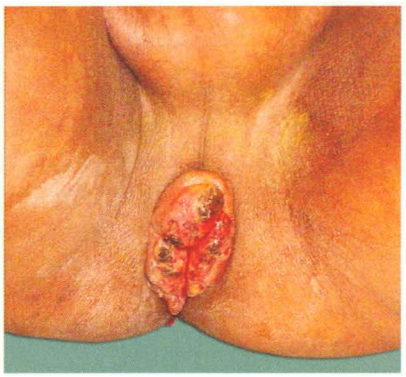


Fig. 31.53: Grade IV

Fig. 31.54: External pile

Fig. 31.55: Prolapsed piles—painful condition

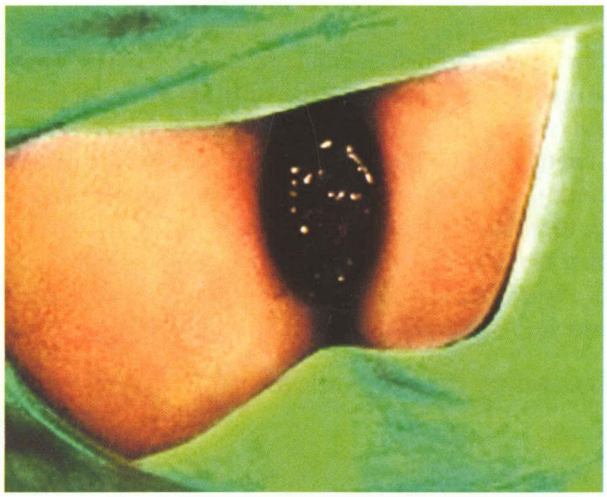


Fig. 31.56: Thrombosed pile mass (*Courtesy:* Dr CG Narasimhan, Senior Consultant Surgeon, Mysore, Karnataka)

- **Sigmoidoscopy and proctoscopy are done to rule out proximal cancer.**

Complications of haemorrhoids (Figs 31.55 to 31.57)

1. It can cause **chronic anaemia**. Rarely, massive bleeding can occur because of portal hypertension.
2. A **prolapse** outside presents with severe pain in the perianal region—piles gripped by internal sphincter results in venous congestion and oedema followed by strangulation. Such patients are treated by:
   - Elevation of foot end of bed
   - Metronidazole 400 mg, 3 times a day for 5 days.
   - Saline dressings to reduce oedema
   - Local lignocaine jelly application
3. **Ulceration** and secondary infection
4. **Thrombosis** and fibrosis

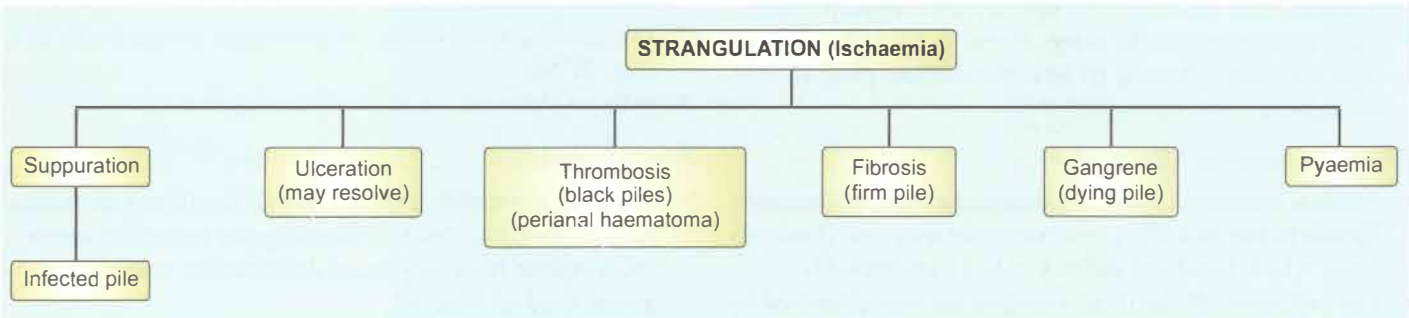


Fig. 31.57: Complications of piles

TREATMENT OF HAEMORRHOIDS

A. Nonoperative treatment: It is indicated in Grade I and Grade II piles which are not causing significant bleeding or discomfort (Key Box 31.13).

KEY BOX 31.13

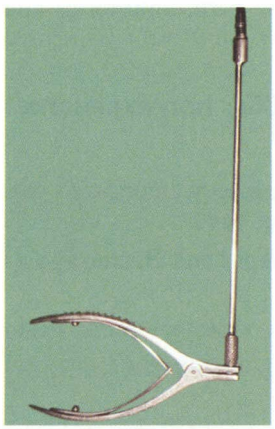
NONOPERATIVE TREATMENT

- Fibre supplementation
- Increased fluid intake
- Bulk purgative—laxatives—isapgul husk, etc.
- Reading in toilet to be discouraged (respond to call and do not strain)
- Encourage to lose weight

Remember as **FIBRE**

B. Injection of sclerosant: 5% phenol in almond oil is injected into **submucosa** above the dentate line. Hence, it is painless. It produces **aseptic thrombosis** of pile mass and is indicated in Grade I. The injection is **perivascular**.

C. Barron's band application: It is indicated for grade II and grade III haemorrhoids, wherein bands are applied at the neck of the haemorrhoids. It causes necrosis and thus, piles get fibrosed. One or two can be banded at a time (Key Box 31.14).



Figs 31.58 and 31.59: Barron's band ligator and band has been applied to one of the pile masses

KEY BOX 31.14

BAND LIGATION: WISDOM LINES

- Bands should be applied **1–2 cm above dentate** line to avoid pain.
- Bands should not be applied in patients who are taking **anticoagulants.**
- Bands **should not be applied for immunocompromised** patients without broad spectrum antibiotics to avoid life-threatening sepsis.
- **Should not band all the three pile** masses at same time. Quadrant by quadrant with a gap of 2 weeks is ideal.
- If severe pain, fever and urinary retention develops after band (sepsis), examine under general anaesthesia and remove band.

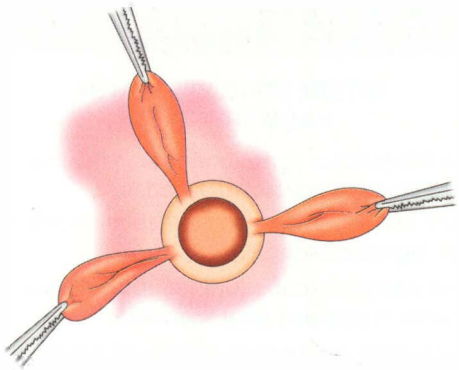


Fig. 31.60: Three pile masses are held with artery foreceps

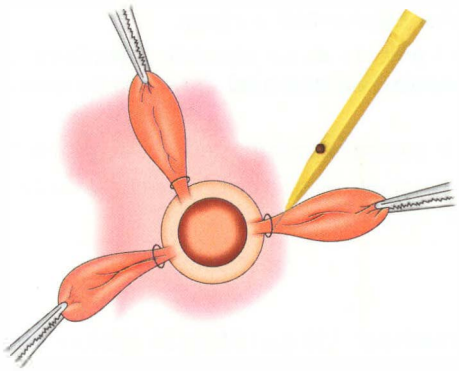


Fig. 31.61: Ligatures are applied at the base of the haemorrhoids and they are being cut with cautery

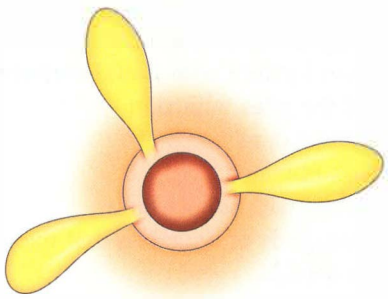


Fig. 31.62: After haemorrhoidectomy the excised portion should look like a clover

D. Operative treatment: Haemorrhoidectomy. Open method, closed method, stapler haemorrhoidopexy.

HAEMORRHOIDECTOMY (Key Box 31.15)

Excision of the pile masses up to base is indicated in Grade II and Grade III hemorrhoids. It can be done by 3 methods: Open, closed and with stapler (*vide infra*).

PEARLS OF WISDOM

Excisional haemorrhoidectomy produces more discomfort (pain) than stapler haemorrhoidopexy but lesser recurrences).

KEY BOX 31.15

INTERESTING WISDOM IN HAEMORRHOIDS

- Haemorrhoids occur due to downward prolapse of vascular cushions into and beyond anal canal.
- Minimum ideal investigation for haemorrhoids should be **flexible sigmoidoscopy.**
- Preserve adequate **mucocutaneous bridges** in excisional procedures to **prevent anal stenosis.**
- **Urgency and tenesmus** following stapled haemorrhoidoscopy responds well to **oral nifedipine.**
- **Metronidazole** is the most important agent in **reducing pain** after haemorrhoid surgery.
- Grade I and II can be injected: **Injections** should be **perivascular, submucosal** and above the level of dentate line.
- Grade III require haemorrhoidectomy or haemorrhoidopexy
- Grade IV require initial conservative treatment followed by surgical procedure.

Types

I. Open method: Milligan-Morgan ligature and excision (Figs 31.60 to 31.62)

- Stretch the sphincter
- Identify the positions of pile masses
- Dissection up to the base (pedicle)
- Transfixation ligature with **nonabsorbable silk**
- Excision of the piles with skin
- Trimming the wound
- Haemostasis obtained
- Wound packed with roller gauze
- A tube drainage is provided so that the blood (oozing) can escape outside.

PEARLS OF WISDOM

Always leave a bridge of skin in between the excised pile masses to prevent anal stenosis.

II. Closed method (Hill-Ferguson)

- Basic steps are the same as above
- Cut mucosa and skin edges are sutured with absorbable catgut sutures.

Postoperative management

1. Strong analgesics, in the form of injection pethidine or morphine, are given to reduce the pain.
2. Antibiotics along with metronidazole are given to prevent secondary infection.
3. Bulk purgatives are given to avoid constipation.
4. **Sitz bath** twice a day is given by using warm saline or $KMnO_4$ solution.

Postoperative complications

They can be classified into early and late complications. Acut retention of urine and haemorrhage are early complication Anal stricture, anal stenosis, anal fissure and incontinence ar the late complications. Few complications are described belov

1. **Retention of urine** is common in men due to severe pair It can be managed by treating the pain and hot wate fomentation in the suprapubic region. Catheterisation i done as a last resort.
2. **Reactionary haemorrhage** is more common. It is due to loose ligature or some opened up bleedings. Generally stop by pressure packing. Otherwise, under anaesthesia, ligat or cauterise bleeding point
3. **Secondary haemorrhage** can occur due to infection. I manifests 6 to 8 days later. If the bleeding is significant exploration in the operation theatre may be necessary. I should be done under anaesthesia. With good illuminatio it is possible to identify the bleeding points and ligate them
4. **Anal stenosis** can occur if too **much skin** is excised during haemorrhoidectomy. It needs regular dilatation.
5. Anal fissure, submucous abscess, and incontinence car occur after haemorrhoidectomy.
6. **Wound infection:** Minor degree of wound infection does occur and can be treated with sitz bath, antibiotics and regular dressings.

STAPLER HAEMORRHOIDOPEXY: non-excisional procedure (Figs 31.63 to 31.68)

- A novel method for 3rd and 4th degree haemorrhoids was introduced by Dr Antonio Longo in 1997.
- It is also called **P**rocedure for **P**rolapse and **H**aemorrhoids (PPH).
- After reduction of prolapsed piles, a prolene purse string suture is applied circumferentially, taking good mucosal bites 3 cm above dentate line.
- This is possible by using **Circular Anal Dilator** (CAD).
- By maintaining traction in the tails of suture, the stapler is fully closed and fired.
- Slowly stapler is opened and withdrawn.
- **Look at 'doughnut'. If it is complete, nothing to worry.**
- Thus a circular ring of mucosal tissue above the level of dentate line is removed. Internal haemorrhoids are not removed, external haemorrhoids are also not removed (eventually they regress).
- Thus, **2 rows of staples and 28 staples** are present.

Advantages

- Lesser operative time
- Less bleeding
- Lesser postoperative pain and need for analgesia
- Lesser postoperative stay at hospital, part of day care procedures

STAPLER HAEMORRHOIDOPEXY

Fig. 31.63: Grade III haemorrhoids before surgery

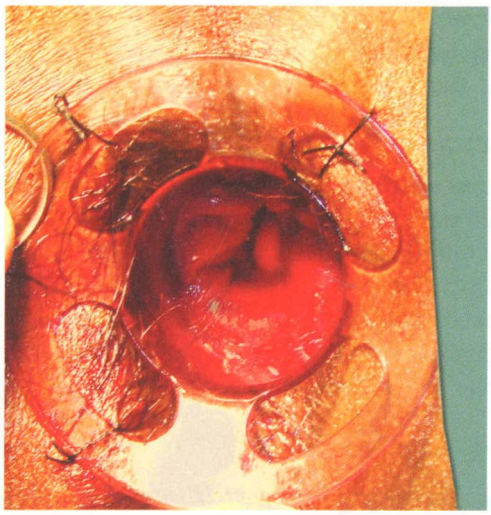


Fig. 31.64: Circular anal dilator is in place

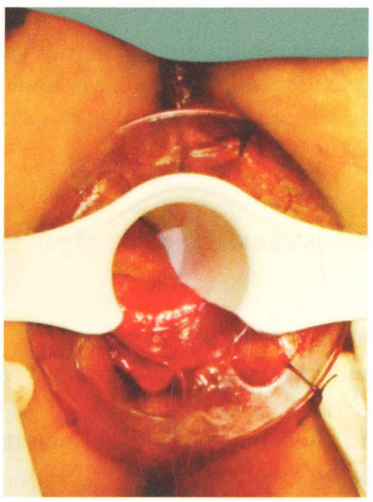
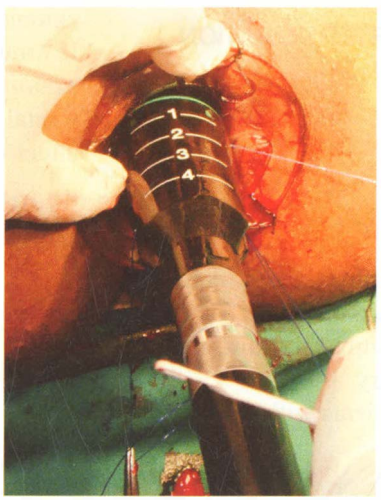


Fig. 31.65: Purse string suture is applied



Figs 31.66 and 31.67: Maintain traction in the tails of purse string suture and stapler is closed and fired

Fig. 31.68: After surgery

Courtesy: Dr BH Anand Rao, Dr Vinayak Shenoy K, Dr Ramachandra L, Dr Hartimath B, Dr Prashanth Shetty, Dr Saritha Kanth (Consultants), and Dr Akshay Nadakarni (Registrar), Department of Surgery, KMC, Manipal.

- Study period: July 2007–2009
- Total number of patients: 186

  93 open, 93 stapled
- Follow up: 2 years

Result of stapled group

- **Advantages:** Lesser postoperative pain, earlier return to normal activities, better satisfaction.
- **Side effects:** Mucoid discharge with tenesmus (5 patients), recurrences in 3 patients.

- Earlier return to normal activities
- No major postoperative short-term and long-term complications
- No long term side-effects such as anal stenosis or chronic pain as may happen with open haemorrhoidectomy.

Disadvantages

- High cost of instrumentation
- Difficult technically and needs special training (learning curve)
- Rare complications such as intra- and postoperative bleeding. Occasional cases of postoperative fissures, mucosal discharge, persistent tenesmus, infection and long-term complications such as rectovaginal fistulae, polyps at stapler line and recurrence.

PEARLS OF WISDOM

In open haemorrhoidectomy: If wound is like clover, it is over. In stapler haemorrhoidectomy: If doughnut is complete, it is time to celebrate.

DOPPLER GUIDED HAEMORRHOIDAL ARTERY LIGATION

- Doppler principle is used to identify the feeding artery to the haemorrhoid mass and it is ligated
- Doppler incorporated proctosope is introduced, once the artery is recognised by audible signal.
- After this, the needle is inserted into the lumen of the proctoscope
- The artery is ligated by **figure of eight** suture. Thus, main artery supplying the haemorrhoid is blocked
- Procedure is simple out patient procedure, no pain, no anaesthesia. No blood loss, early recovery
- Safe in all types of patients including patients with serious morbidity

External haemorrhoids

- Described by Milligan as **5 day painful self-curing lesion.**
- Constipation and sudden straining at stools or lifting weights will result in a tender subcutaneous swelling at the anal margin.
- It is bluish in colour because it is a thrombosed vein or venule (external haemorrhoid).
- If tenderness is extreme, under local anaesthesia, incision can be given over the swelling and clot can be evacuated.
- In other cases, it will resolve within 5 days after fibrosis/ suppuration.

ANORECTAL ABSCESS

Acute anorectal suppuration—anorectal abscess

- More common in men especially diabetic. Blood-borne infection is common in diabetic patients.
- Mostly originate from the anal gland opening at the base of the anal crypts. This is **cryptoglandular theory of intersphincteric anal gland infection** described by Sir Allan Parks. From here, pus spreads along path of least resistance—thus form perianal abscess or ischiorectal abscess (Key Box 31.16).
- Other source of anorectal sepsis is foreign body, trauma, sexually transmitted diseases for lower level abscesses. Crohn's disease and carcinoma rectum with perforation may form pelvirectal abscess (supralevator).
- Typically patients presents with high grade fever with chills and rigors. On examination, a tender indurated swelling is found in the perianal region or in the ischiorectal fossa.
- Culture usually shows *E. coli* in about 70–80% of cases.
- *Staphylococcus aureus,* Streptococcus, Bacteroides are the other organisms.

Types (Fig. 31.69)

1. Perianal abscess

- It occurs due to infection of anal glands in the perianal region.
- It may be due to a boil, anal gland infection or thrombosed external pile.
- It produces severe pain, throbbing in nature and on examination a soft, tender, warm swelling is found.
- Rectal examination reveals a tender, boggy, swelling under the anal mucosa.

Treatment

Antibiotics, incision and drainage and excision of part of skin (roof).

2. Submucous abscess

- Collection of pus under the mucous membrane of rectum or anal canal.
- It can also be due to infection of injected haemorrhoids. It can be drained using proctoscope.

3. Ischiorectal abscess

- Collection of pus in the ischiorectal fossa, which is lateral to rectum and medial to pelvic wall.
- Bounded above by levator ani and inferiorly by pad of fat in the ischiorectal fossa.
- Ischiorectal fat is poorly vascularised. Hence, it is more vulnerable to infection.
- Abscess occurs due to spread of perianal abscess or due to blood-borne infection.
- Diabetes is the precipitating factor

KEY BOX 31.16

CAUSES OF ANORECTAL ABSCESS

- Infection
- Irritation (Crohn's disease, ulcerative colitis)
- Immunity low (diabetes, AIDS)

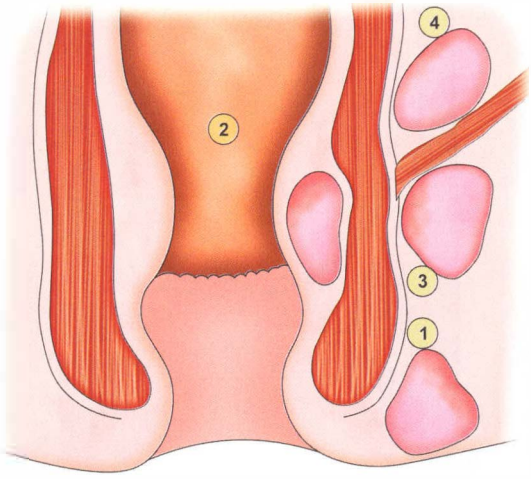


Fig. 31.69: Types of anorectal abscess (*see* text)

Clinical features

- Severe throbbing pain is characteristic of ischiorectal abscess.
- Induration in the ischiorectal fossa
- Common in diabetic men
- Frank evidence of abscess such as fluctuation need not be seen and is a late sign (Key Box 31.17).
- High grade fever with chills and rigors.
- Per rectal examination is painful and bogginess can be appreciated on the side of the lesion.

Treatment

Under anaesthesia, a cruciate incision (+) (Figs 31.70 and 31.71) is made and the 4 flaps are raised. All the pus is evacuated and the wound is packed with iodine roller gauze and left open. Edges of the skin are trimmed to leave an opening so that drainage of pus occurs freely. It heals with granulation tissue within 10–15 days. Appropriate antibiotics are given for a period of 5 to 10 days.

4. Pelvirectal abscess

It is a pelvic abscess, which is drained through the rectum. The common causes are pelvic peritonitis, appendicitis, septic abortions, etc. The details of the causes, clinical features and the management are discussed in page 647.

KEY BOX 31.17

DEEP ABSCESS WITHOUT FLUCTUATION

- Ischiorectal abscess
- Breast abscess
- Parotid abscess
- Prostatic abscess
- Midpalmar abscess

Fig. 31.70: Aspiration of ischiorectal abscess

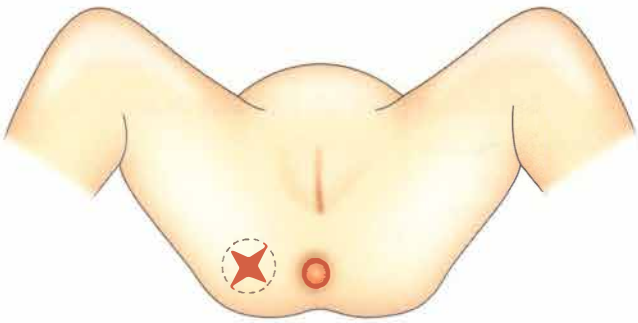


Fig. 31.71: Cruciate incision—drainage

See Key Box 31.18 for summary

KEY BOX 31.18

ANORECTAL ABSCESS

- Common causes such as boil or infected sebaceous cyst has to be ruled out first.
- Remember other causes such as infection following haemorrhoidal injection or band ligation.
- Uncommon causes such as foreign body or penetrating trauma also can give rise to anorectal abscess.
- **Last but not the least, AIDS and diabetes have to be ruled out in all cases of anorectal abscess.**

FISTULA *IN ANO*

Abnormal communication between anal canal and rectum with exterior (perianal skin) is called as fistula *in ano*. Even though multiple openings are seen in the perianal skin, **the internal opening is always single**.

Aetiopathogenesis

1. They occur due to **persistent anal gland infection,** which results in anorectal abscesses, rupture inside as well as outside resulting in a fistula. Once a fistula occurs, it persists because of infection and absence of rest to the part. As there are many anal glands, often, problem persists inspite of initial treatment of one fistula.

2. In India, **tuberculosis** is common. Patients with pulmonary tuberculosis have 1–2% chances of developing multiple anal fistulae. Whenever a patient presents with multiple anal fistulae, it is but natural to think of tubercular aetiology. Such fistulae are **not indurated and there is watery discharge without pus.**
3. In Western countries, **ulcerative colitis and Crohn's disease** are responsible for multiple anal fistulae.
4. **Colloid carcinoma of rectum** can present as multiple fistulae *in ano*. This type of carcinoma has worst prognosis. Rectal examination should be done in every patient with anal fistula.

- Other causes of anal fistula (Key Box 31.19)

KEY BOX 31.19

SPECIAL TYPES OF FISTULA *IN ANO*

- **F**istula carcinoma
- **I**leitis—Crohn's
- **S**chistosomiasis
- **T**uberculosis
- **U**lcerative colitis
- **L**ymphogranuloma venereum
- **A**nal fissure abscess

Students can remember as **FISTULA**

Classification

1. Standard classification (Fig. 31.72)

1. Subcutaneous
2. Submucous
3. Low anal
4. High anal
5. Pelvirectal

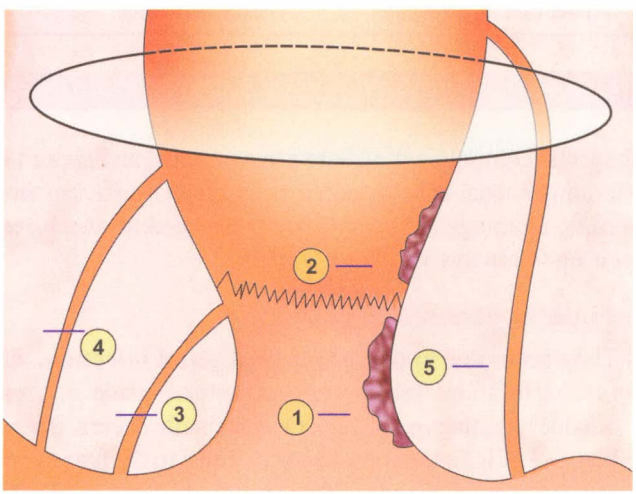


Fig. 31.72: Standard classification (*see* text)

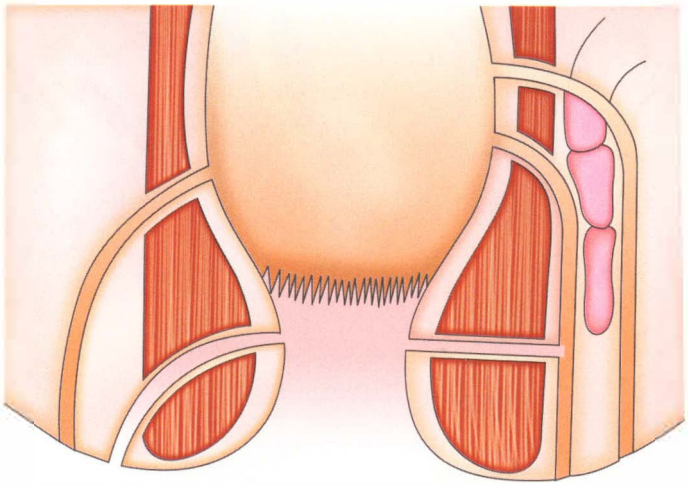


Fig. 31.73: Park's classification

II. Park's classification (Fig. 31.73)

1. Intersphincteric
2. Trans-sphincteric
3. Supralevator (internal opening is situated above the anorectal bundle).

Clinical features

- Persistent seropurulent discharge, keeps the part always wet.
- Previous history of anal gland infection, with recurrent abscess.
- External opening can be single/multiple, with pouting granulation tissue, may discharge blood.
- Internal opening in carcinoma felt as a 'button hole' defect inside the rectum.
- **Goodsall's rule:** A fistula, with an external opening in the anterior half of anus within 3.75 cm tends to be direct type and in the posterior half, indirect type or curved and sometimes horseshoe type. It may communicate with the opposite side (Fig. 31.74).

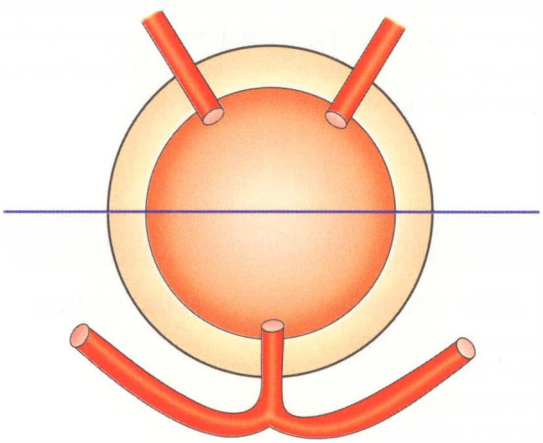


Fig. 31.74: Goodsall's rule

Diagnosis

- External opening is found at the bottom of a depressed area or with granulation tissue or it is seen discharging pus.
- Internal opening may be felt on digital examination as **indurated** area or sometimes can be seen with proctoscopy or after sigmoidoscopy.
- The entire track may be palpable as indurated cord like structure.
- **Endorectal ultrasonography** and **MRI** seem to identify internal openings and fistula. However, they can be selectively used in deserving cases.
- Examination under general or regional anaesthesia.

TREATMENT (Figs 31.75 to 31.80)

I. Fistulotomy

It is indicated in low fistula (internal opening below the anorectal bundle). A probe is passed through the external opening into the rectum and along the length of this tract the fistula is laid open. It is done under anaesthesia. The wound is left open and allowed to heal by granulation tissue developing from the floor of the fistula (marsupialisation).

Intersphincteric and low trans-sphincteric fistulas of recent origin are treated by fistulotomy and marsupialisation.

Advantages

- Least chances of recurrence
- Relatively easy procedure
- Minor degree of incontinence.

II. Fistulectomy

All **chronic fistulae (low) are treated by fistulectomy** by excising the entire fibrous tissues and tract. Here also, the wound is kept open. This can also be done for **posterior semi horseshoe and horseshoe** fistula. Some incontinence can occur.

Fig. 31.75: Multiple recurrent fistula *in ano*—biopsy reported as tuberculosis

Fig. 31.76: Fistula—rectovaginal fistula

III. Fistulectomy with or without colostomy

It is indicated in high fistula *in ano*. The internal opening is situated above the anorectal bundle. Hence, during fistulectomy, there is a chance of injury to the anorectal bundle and may cause incontinence. Temporary or permanent colostomy may be necessary. If there is a cause, treat the cause. Surgery of intersphincteric fistula and trans-sphincteric fistula may result in incontinence.

IV. Use of seton or medicated thread (Ksharsutra)

Ksharsutra is an ayurvedic term. It is a medicated thread passed through the entire tract and both ends are tied and tightened once a week so that by 6 weeks it cuts through (Key Box 31.20).

Fig. 31.77: Low fistula *in ano*

Fig. 31.78: Fistulotomy by passing a fistula probe

Fig. 31.79: Fistula *in ano*—excision is going on

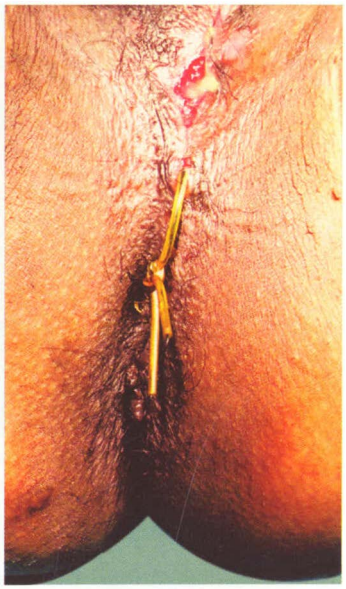


Fig. 31.80: Fistula *in ano*, seton in place

KEY BOX 31.20

SETON

- It is a Latin word. Seton means bristle—material such as thread, wire, or gauze that is passed through subcutaneous tissues.
- Varieties of materials used as setons—plastic tubes, infant feeding tubes, proline suture material, medicated thread used in Ayurvedic method—Ksharasutra. **Ksharsutra** is a sanskrit phrase in which *Kshar* refers to anything that is corrosive or caustic; while *sutra* means a thread.
- Loose setons for long-term palliation. Examples: Fistulae associated with Crohn's disease, complicated recurrent fistulae.
- Cutting tight setons: Used in complicated high fistulae wherein a fistulotomy may result in anal incontinence. So, seton is tied, patient will tighten it everyday for a period of 8–12 weeks till it comes down. Once it comes down, seton is removed. This will decrease the chances of incontinence.
- Main advantage of seton is it eliminates sepsis by keeping the track open.
- Disadvantage is that patient will always feel a foreign body sensation in the rectum and anal canal.

See Key Box 31.21 for recent advances in fistula surgery.
See Ten commandments for fistula *in ano*.

FISSURE *IN ANO*

Definition

Longitudinal tear in the lower end of anal canal results in fissure *in ano*. It is the **most painful condition** affecting the anal region. Commonly seen in young patients.

KEY BOX 31.21

RECENT ADVANCES IN FISTULA SURGERY

1. **Biological agents:** The basic principle is to plug and seal the tract. It allows the ingrowth of healthy tissue. Thus initially fibrin glue was used but results are not good in long term. Porcine small intestinal mucosa or porcine dermal collagen also has been used. Results are not satisfactory.
2. **Video-assisted anal fistula treatment** (VAAFT): A novel sphincter-saving procedure for treating complex anal fistulas (more details in page 1179).
   - **V**isualisation of the fistula tract using the fistuloscope
   - **A**im is correct localisation of the internal fistula opening under direct vision
   - **A** stapler or cutaneous-mucosal flap to close the internal opening after endoscopic treatment of the fistula.
   - **F**istuloscopy is done under irrigation and followed by an operative phase of fulguration of the fistula tract
   - **T**otal closure of the internal opening and suture reinforcement with cyanoacrylate.

   You can remember as **VAAFT**

TEN COMMANDMENTS FOR FISTULA *IN ANO*

1. Should find out the internal and external openings
2. Should try to define the type of the fistula in relation to sphincter
3. Should define low or high fistula
4. Should rule out special types of fistula
5. Should conduct thorough examination again under anaesthesia before surgical procedure
6. Should do MRI in difficult, recurrent and complicated fistula
7. Should do fistulotomy in all intersphincteric fistulae and trans sphincteric involving 30% of the voluntary musculature
8. Should do fistulectomy in low fistula—it will open up the infected cavity better even though wound will be bigger than a simple fistulotomy wound
9. Should use setons in high fistula or complicated fistula wherein a fistulotomy may result in recurrence or incontinence or when staged procedures are planned
10. Should explain to the patient about possibility of some degree of incontinence and take consent for colostomy in high fistula

Aetiopathogenesis (Fig. 31.81 and Key Box 31.22)

- 90% of anal fissures occur in the **posterior part of anal** canal and 10% anteriorly. It is initiated by hard stool causing a crack. As a result of this, defaecation results in pain. Anal fissure is more common posteriorly in the midline because of relative ischaemia.
- **Due to pain, internal sphincter spasm** takes place which makes constipation worse resulting in a **chronic fissure**.
- **Anterior fissures** occur in elderly women secondary to repeated pregnancies. This is due to damaged pelvic floor

and lack of support to anal mucous membrane. Acute fissure in females may occur after vaginal delivery.

PEARLS OF WISDOM

Fissure away from midline should raise the possibility of Crohn's disease, sexually transmitted diseases, etc.

KEY BOX 31.22

VARIOUS FACTORS WHICH PRECIPITATE ANAL FISSURE

- **F**aeces—hard
- **I**schaemia
- **S**urgical procedures—haemorrhoidectomy
- **S**phincter hypertonia
- **U**nderlying diseases—Crohn's, sexually transmitted diseases, etc.
- **R**epeated childbirth
- **E**nthusiastic usage of ointments and abuse of laxatives

Remember as **FISSURE**

Clinical features

- Severe pain during and after defaecation, burning in nature, lasting for about ½ to 1 hour because of which defaecation is postponed.
- Severe constipation is present.
- Stools are hard, pellet like and there is a drop of blood or streaks of fresh blood.

PEARLS OF WISDOM

Drop of blood is due to anal fissure. Splash of blood is due to haemorrhoids, bloody slime is due to carcinoma.

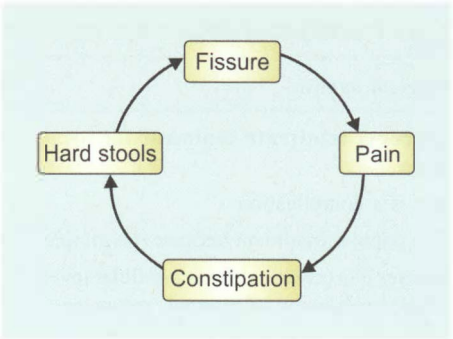


Fig. 31.81: Aetiopathogenesis of anal fissure

- Sentinel pile refers to tag of skin at the outer end of the fissure.
- In some cases, fissure may be associated with a small perianal abscess resulting in worsening of pain.

Diagnosis (Table 31.5)

1. When the buttocks are spread apart, a longitudinal tear and a hypertrophied, thickened skin is seen near the lower end of fissure—**sentinel pile**.
2. Per rectal examination can be done (with lignocaine jelly application) and **sphincter spasm** can be appreciated.
3. Proctoscopy is contraindicated because the condition is very **painful**.

Treatment (Table 31.6)

I. Conservative

- Avoid constipation—encourage fibre diet, mild laxatives and not to postpone defaecation.
- Surface anaesthetic creams: Lignocaine jelly.
- Metronidazole and antibiotics
- Sitz bath

Table 31.5 Difference between acute fissure *in ano* and chronic fissure *in ano*

Acute	Chronic
• Sudden onset—example after vaginal delivery or following hard stools • Acute pain in the anal canal, severe burning after defaecation with bleeding • No itching around anal opening • Severe sphincter spasm, small crack in the lower anal canal • No sentinel pile—tag of skin • PR very painful • Proctoscopes—better not try insertion • Usually responds to conservative treatment—local application of glyceryl trinitrate (GTN) 0.2% 3–4 times a day or diltiazem 2% twice a day • Emergency sphincterotomy may be required in a few patients	• A few months duration of symptoms • Chronic pain in the anal canal, burning after defaecation with bleeding—few exacerbations • Itching is usually present due to ulcer or hypertrophied skin-sentinel pile • Sphincter spasm, chronic canoe shaped ulcer in the lower anal canal • Sentinel pile—tag of skin • PR painful • Proctoscope—can be done with proper ligonocaine application to the anal canal • Responds to conservative treatment but effect is temporary surgery is the treatment of choice • Lateral sphincterotomy or other procedures may be required

Table 31.6 Treatment of chronic fissure *in ano*

Pharmacological agents	Injection botulinum A toxin	Lateral sphincterotomy/flaps
• 0.2% **glyceryl trinitrate** ointment for local application • Headache is a complication • Still it is a popular treatment because it is simple • Drug releases NO (nitric oxide) at a cellular level and mediates relaxation of internal sphincter • **Oral nifedipine** 20 mg, twice daily. Topical nifedipine also helps	• Healing rate is 80% in chronic anal fissures • 6% recurrence in 6 months • Single injection, simple method, smooth recovery—chances of sepsis are present	• Gold standard • Manometry of anal sphincter can be done before procedure especially in women because 30–40% of patients develop incontinence following lateral sphincterotomy **Advancement flap** Women—postpartum fissure—poor anal tone

II. Agents which decrease sphincter pressure

- **Glyceryl trinitrate** (0.2%) topical application: Significant headache and 50% recurrence are drawbacks. It reduces spasm, increases vascular perfusion.
- **Purified botulinum toxin** injection into internal spincter: It inhibits presynaptic release of acetylcholine from cholinergic nerve endings and cause temporary paresis of striated muscle. Cost, perianal thrombosis and sepsis are drawbacks. Injection produces **prolonged but reversible effects,** thus avoiding **permanent injury** (Key Box 31.23).
- **Calcium channel blockers:** Nifedipine, diltiazem oral and topical applications **(2%)** also have been used.

KEY BOX 31.23

ROLE OF BOTULINUM TOXIN INJECTION

- Achalasia cardia and other oesophageal motility disorders.
- Anal fissures
- Sphincter of Oddi dysfunction
- Frey syndrome

III. Surgical treatment

1. **Lateral anal sphincterotomy of Notaras** (or dorsal) is **the best alternative procedure**. Here internal sphincter is divided away from fissure either in right or left lateral positions. The procedure can be easily done by using a bivalved speculum in the anal canal. This is the procedure of choice. **Sphincterotomy should be limited to the length of fissure to avoid incontinence.**
2. **Fissurectomy and local advancement flap:** This is indicated in **persistent, chronic, nonhealing fissure.** After excision of the fissure, the resulting defect in the anal canal is closed by a small (**rhomboid**) advancement flap. This should be considered not as a first line of treatment. Recovery from this operation takes much more time than other treatments for anal fissures.
3. **Lord's dilatation:** It is also called **blunt sphincterotomy**—few fibres of internal sphincter are divided. It relieves the spasm and the fissure heals. Rarely, in female patients it may result in incontinence. **It is not a recommended treatment nowadays.**

PEARLS OF WISDOM

Lateral sphincterotomy is very popular and gives good results.

PILONIDAL SINUS (JEEP-BOTTOM)

- Pilonidal sinus means **nest of hairs** in Greek. Also called **Jeep-bottom** because it was very common in jeep drivers.
- More common in dark people than fair people.
- It is an acquired condition, commonly found in hairy males. **It is acquired due to following reasons**
  - Appears between the age of 20 and 30 years
  - Hairy men are more affected
  - The **hair follicle is never demonstrated in the wall of the pilonidal sinus** but hair is the content of pilonidal sinus.
- Hair accumulates due to vibration and friction causing shedding of the hair. Thus, it accumulates in the gluteal cleft and enters the opening of the sweat glands.
- Pointed end of the dead hair is inside (blind end of the sinus).

Clinical features

- External opening of the sinus seen just above the anal verge in the midline over the coccyx (Key Box 31.24).
- History of discharge of pus
- History of recurrent abscesses which rupture, discharging pus.
- Can be asymptomatic

Diagnosis

Osteomyelitis of the coccyx is the only differential diagnosis for pilonidal sinus. Hence, X-ray of the coccyx should be taken.

Treatment (Figs 31.82 to 31.85)

- Inject methylene blue to demonstrate branches of the sinus followed by **excision of the sinus**. The patient is positioned prone with buttocks elevated (**Jack knife position**).

KEY BOX 31.24

SITES OF PILONIDAL SINUS

- Midline over the coccyx
- Umbilicus
- Interdigital in barbers

- After excision there are two methods to treat the wound—Open and closed methods (Key Box 31.23).
- **Open method:** The wound is left open after excision followed by regular packing with iodine or eusol gauze pieces (Key Box 31.25).
- It may take 3–4 weeks for the healing of pilonidal sinus. Regular sitz bath is also given.
- This method carries the least recurrence.
- **Closed method:** The wound is closed by 'z' plasty. This method carries 10–20% chances of recurrence. Rhomboid flap (Limberg flap) can be raised to close the defect also.
- **Karydakis procedure:** Primary procedure is to remove all the sinus tracts and their branches till sacral bone. In this operation, semilateral incision is made around the sinuses and flaps mobilised to excise all the sinuses and their branches. Then tension free closure is done. Compared to elliptical incision, this incision and closure has decreased chances of skin necrosis.
- **Bascom's technique:** In this procedure, an incision is given laterally, not in the midline. After raising the flaps, wide excision of the infected sinuses and tracts is done followed by closure of the midline openings. Lateral wound is left open (in the conventional operation, midline wound is left open).

PEARLS OF WISDOM

Very, very rarely carcinoma can arise in a chronic pilonidal sinus.

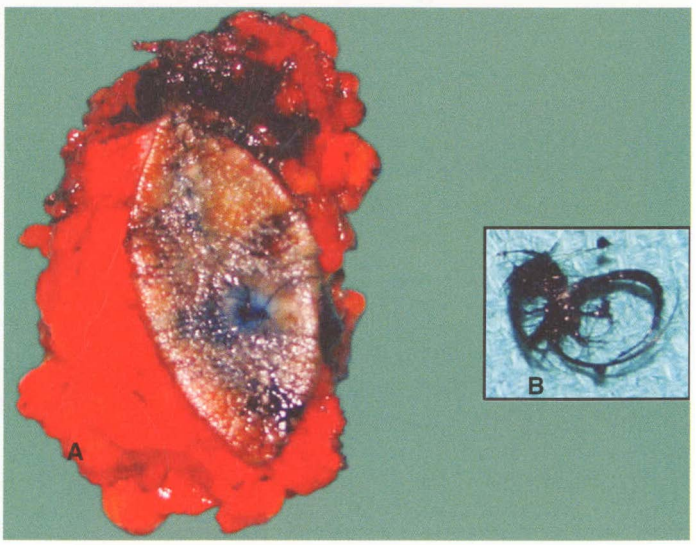
KEY BOX 31.25

PILONIDAL SINUS

- It is acquired condition—popularly called Jeep-bottom.
- Common in hairy men
- Multiple sinuses communicating with each other
- They open to the exterior by multiple openings
- The direction of the sinuses is cephaloid
- Recurrent abscesses which rupture are common
- Excision with or without marsupialisation, flap closure or z plasty are the treatment options.
- Inspite of the adequate surgical procedures, recurrence is common.

Fig. 31.82: A case of pilonidal sinus—methylene blue is first injected into the opening. The excision area is marked also with methylene blue

Fig. 31.83: Blue stained tissues are dissected all around and removed



Figs 31.84A and B: (A) Specimen of excised tissue with sinus track, and (B) shows tuft of hair which was present within the sinus

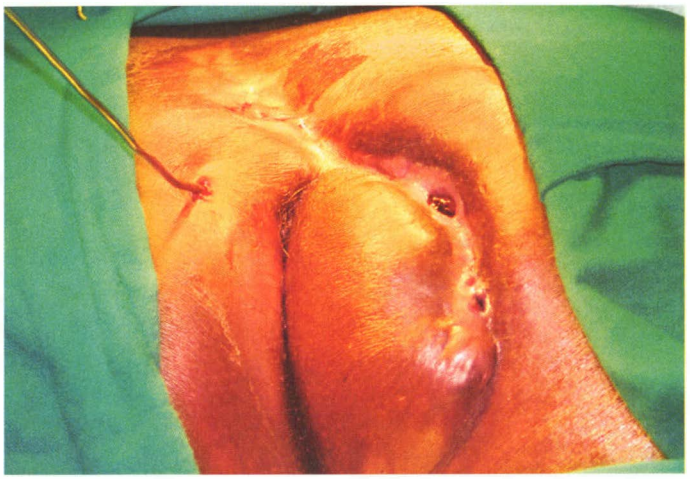


Fig. 31.85: Neglected, persistent, recurrent pilonidal sinus. Fistula probe is introduced into one of the openings followed later by demarcation of the fistula

SACROCOCCYGEAL TERATOMA

- It is a congenital condition affecting the sacrococcygeal region.
- In this region, totipotential cells persist for a longer period compared to rest of the area. Hence, it is the site of teratomas.

Clinical features

- 20% of the cases are stillborn babies. It is common in a female child.
- Presents as a swelling in the sacrococcygeal region pushing the rectum anteriorly.
- The surface of the swelling ulcerates. Many cystic areas are present in the swelling.
- The swelling is fixed to the sacrum and coccyx from which it is impossible to separate/isolate.

Complications

- Ulceration
- Secondary infection
- Haemorrhage
- Teratocarcinomatous change occurs by one year of age.

Treatment

Excision of the teratoma with part of sacrum and coccyx.

MALIGNANT TUMOURS OF ANAL CANAL

- They are not uncommon tumours which present with bleeding per rectum, burning and itching in the anal region.
- The diagnosis is obvious in many cases once buttocks are separated or by digital examination.
- Tissue diagnosis is a must before radical treatment.

Types

1. **Squamous cell carcinoma:** Papillomas are the chief predisposing factors. Local excision or APR (abdomino perineal resection (APR) is the treatment with external RT in appropriate cases (Fig. 31.86).
   - For sphincter preservation—chemoradiation can be used. It is called Nigri's regime.
2. **Basaloid carcinoma:** It is a highly malignant, non-keratinising, squamous cell carcinoma. Treatment is similar to squamous cell carcinoma.

PEARLS OF WISDOM

Basal cell carcinoma is very rare in the anal canal.

3. **Melanoma:** Beware of a patient who comes with bilateral groin nodes which are bulky. The patient may be having

Fig. 31.86: Squamous cell carcinoma anal canal

Fig. 31.87: Melanoma anal region

Fig. 31.88: Melanoma resembled prolapsed piles but pigmentation was evident on closer examination

KEY BOX 31.26

ANAL INTRAEPITHELIAL NEOPLASIA (AIN—BOWEN'S DISEASE)

- It is squamous cell carcinoma *in situ* of the anus.
- It is precursor to an invasive squamous cell carcinoma.
- It is associated with human papilloma virus type 16 and 18 (HPV 16,18).
- Anoscopy, biopsy to be done
- Dysplasia is an indication for resection/ablation.

malignant melanoma of anal canal—bluish/blackish ulcer in the anal canal. APR is potentially curable in early cases of melanoma. If metastasis is present, the prognosis is poor. So, only local excision is done so as to provide palliation but colostomy is avoided (Figs 31.87 to 31.90).

4. **Adenocarcinoma** is rare. It can occur from the anal glands in pre-existing anal fistula. APR with 5-FU and radiation therapy is indicated.

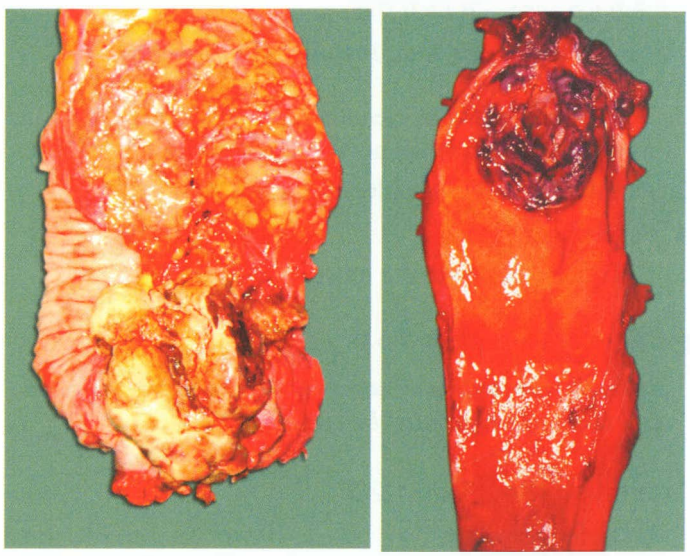
PEARLS OF WISDOM

Please note that Bowen's disease, Paget's disease or verrucous carcinoma, squamous and basal cell carcinoma can also occur in the skin of anal margin.

STRICTURE OF ANAL CANAL AND RECTUM

Causes

1. **Postoperative:** Haemorrhoidectomy, pull-through operations, repeated diathermy fulguration of polyps.
2. **Irradiation**: It occurs one to two years after irradiation.
3. **Senile strictures**
4. **Lymphogranuloma inguinale**: A sexually transmitted disease affecting both male and female patients. Initially pararectal lymph nodes are enlarged followed by development of multiple rectal strictures.
5. **Inflammatory bowel diseases**: Both ulcerative colitis and Crohn's disease result in rectal strictures (5–10%).
6. **Rare**: Congenital, amoeboma, carcinoid, endometriosis, tuberculosis, CMV colitis (Figs 31.91 and 31.92).



Figs 31.89 and 31.90: Malignant melanoma of the anal canal—APR specimens

Clinical features

Increasing constipation is the characteristic feature of stricture of the rectum. It may be associated with hard stools, bleeding and pain in some cases. Per abdominal examination may reveal loaded colon with scybalous masses. Rectal examination can detect a stricture.

PEARLS OF WISDOM

It is mandatory to rule out carcinoma rectum which is the most common cause of stricture.

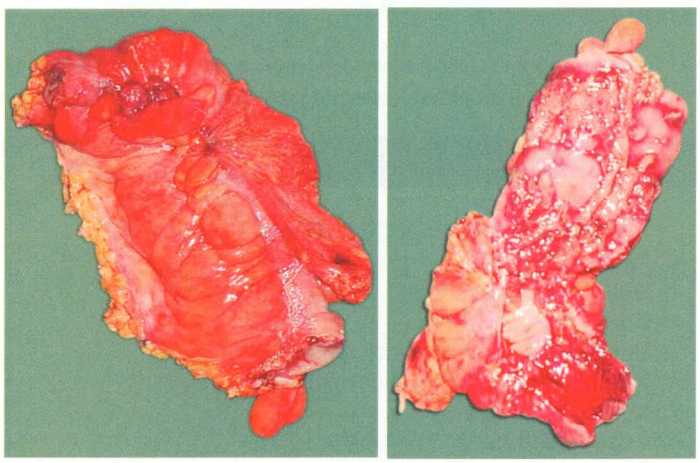
Treatment

1. Conservative treatment includes bulk purgatives, a vegetable diet.
2. Regular dilatation may be necessary for the strictures situated low in the rectum and anal canal.
3. Intractable strictures need to be resected.
4. Treatment of the primary disease.

ANAL INCONTINENCE

Mechanism of anal continence

- Distension of rectum causes **tonic contraction of external sphincter**. This is controlled by cerebrum and the centre is in the **lumbosacral region of the spinal cord.**
- Faeces in contact with anal canal stimulates the specialised nerve endings. Nerve endings are also present in the puborectalis.



Figs 31.91 and 31.92: Rectal stricture due to CMV colitis (*Courtesy:* Dr Satyanarayana N, Dr Srinivas Pai, Dr Madhu, KMC, Manipal)

- **High pressure** in the anal canal (25–120 mmHg) and angle between rectum and anal canal (80°) are the important factors which maintain anal continence.

Anorectal ring

- It marks the junction between the rectum and anal canal.
- It is formed by puborectalis, highest part of internal sphincter, longitudinal muscle and external part of sphincter.

Causes of anal incontinence (Key Box 31.27)

1. **Traumatic:** Injury to the anorectum due to sharp penetrating objects occurs due to accidents.
2. **Surgical procedures:**
   - Damage to the internal and external sphincter can occur due to Lord's dilatation*, a procedure done for fissure *in ano*. However, most of it is temporary.
   - Division of high fistula *in ano* may result in incontinence.
   - Following pull-through procedures done for anorectal anomalies, Hirschsprung's disease.
   - Haemorrhoidectomy—very large pile masses.
   - Extensive small bowel resection
   - Rectal excision
3. **Mass in the anorectum:** Prolapse piles, prolapse rectum and carcinoma rectum may produce temporary incontinence which subsides after surgical procedures.
4. **Neurological causes:** In females, **pudendal nerve neuropathy** which occurs due to chronic straining may result in incontinence. Spinal injuries, spina bifida, meningomyelocoele are associated with anal incontinence.
5. **GI motility increase:** Inflammatory bowel diseases irritate bowel and produce temporary incontinence.
6. **Childhood/congenital causes:** Anorectal malformations, Hirschsprung's disease, spinal bifida, abnormal behaviour.
7. **Miscellaneous:** Old age (senility), general debility and faecal impaction Parkinson's disease, behavioral problem, etc.

Treatment

I. **Temporary incontinence:** Reassurance. Perineal exercises to improve the tone of internal and external sphincter.

KEY BOX 31.27

COMMON CAUSES OF ANAL INCONTINENCE

- **T**rauma
- **R**epeated pregnancies
- **A**nal surgery
- **U**nnatural sex—anal intercourse
- **M**egacolon—Congenital or acquired
- **A**geing or senility

You can remember as **TRAUMA**

*Lord's dilatation or blunt sphincterotomy is no longer done

II. **Permanent incontinence**

1. Divided sphincter can be reunited, followed by overlapping of the remaining muscles.
2. Intersphincteric repair of puborectalis sling and plication of the external sphincter.
3. Gracilis muscle can be used to create a new anal sphincter by transposing it followed by electrical stimulation using a pacemaker.
4. Using artificial sphincter.

PROCTALGIA FUGAX

- This condition is characterised by attacks of severe cramp-like pain arising in the rectum.
- Anxiety status, straining at stools or ejaculation are a few precipitating factors.
- The pain may be unbearable, may recur at irregular intervals. It is possibly due to segmental cramp in the pubococcygeus muscle. The pain usually lasts for a few minutes and subsides (fleeting perianal pain).
- Symptomatic treatment in the form of analgesics are given.

PRURITUS ANI

Definition

This is intractable itching around the anus.

Causes

Perianal and anal discharge: Anal fissure, fistula *in ano*, prolapsed piles, polyps, genital warts are a few conditions which render the anus moist.

PEARLS OF WISDOM

Mucous discharge is an intense pruritic agent.

2. Poor hygiene, lack of cleanliness, excessive sweating and wearing tight and rough underclothing are common causes.
3. Parasitic causes—threadworms
4. Psychoneurosis
5. Allergy, diabetes are the other causes.

PEARLS OF WISDOM

Sexually transmitted diseases such as herpes, anal warts and HIV infection must be excluded.

Treatment (Key Box 31.28)

- Hygienic measures
- Prednisolone topical cream 1% with antifungal agent (miconazole nitrate 2%)

KEY BOX 31.28

PRURITUS ANI—AVOID

- Toilet paper
- Soap
- Too tight underclothing
- Too many ointments
- Local anaesthetic cream

- Moisturising cream/lotion
- Antihistamine—promethazine hydrochloride 10–25 mg at night times.

PEARLS OF WISDOM

Pelvic floor dysfunction also referred to as nonrelaxing puborectalis syndrome is called as ANISMUS. These patients present with constipation. It is a difficult problem to treat.

HIDRADENITIS SUPPURATICA

Definition

It is a chronic recurrent suppuration of apocrine glands in the skin resulting in multiple abscesses which rupture causing multiple sinuses.

Sites

Axilla, groin, back, buttocks and anal regions are common sites.

Pathogenesis

- Occlusion of the gland ducts result in stasis, bacterial proliferation, abscess, rupture. Common organisms are *Staphylococcus aureus* and anaerobes (somewhat like breast abscess).
- Anogenital disease is more common in men hence androgens may play a role in this condition.
- Obesity is another contributing factor.

Clinical features

- Common after puberty till the age of 40 years.
- Typically it is a folliculitis presenting as multiple boils which are painful.
- Pus formation, rupture and persisting sinuses are common
- Interestingly, it does not affect above the level of dentate line nor sphincters.

Differential diagnosis

All diseases resulting in multiple sinuses in and around perineum are the differential diagnosis such as Crohn's disease, tuberculosis, lymphogranuloma venereum, pilonidal sinuses, actinomycosis, etc.

Treatment

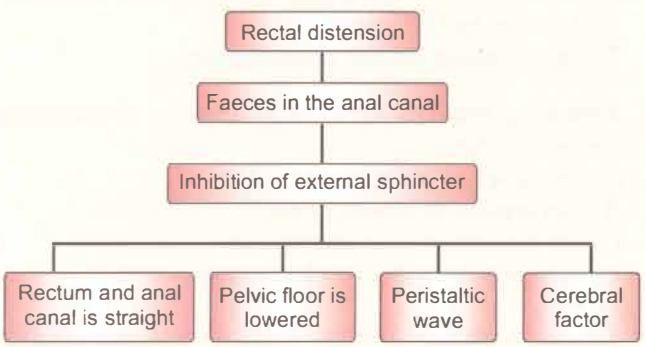
- When in doubt, rule out other causes mentioned above and if necessary a good biopsy from the sinus tract and from the edge of the sinus.
- General measures such as weight reduction, antibiotics, antiseptic medicated soaps, washing the part with warm saline or water.
- Surgery includes laying open all the openings or wide excision with or without skin (radical excision) and direct closure or skin grafting of flap reconstruction are the other choices.

MISCELLANEOUS

Act of defaecation (Key Box 31.29)

KEY BOX 31.29

ACT OF DEFAECATION



WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been upgraded.
- Pelvic floor anatomy and more details about prolapse rectum have been added.
- Haemorrhoidal artery ligation has been added.
- VAAFT has been added

MULTIPLE CHOICE QUESTIONS

1. Splash in the pan is classically described for bleeding from which condition ?
A. Carcinoma rectum
B. Fissure *in ano*
C. Haemorrhoids
D. Polyp

2. Which of the following are causes of anorectal fistulae in males *except:*
A. Crohn's disease
B. Tuberculosis
C. Ulcerative colitis
D. Lymphogranuloma venereum

3. Following are true about peritoneal coverings/fascia of the rectum *except:*
A. Upper one-third is completely covered
B. Middle one-third is covered anterolaterally
C. Lower one-third is covered anteriorly
D. Facia Waldeyer's separates the rectum from sacrum

4. About signet ring carcinoma rectum following are true *except:*
A. It is seen in young patients
B. Cells are filled with mucus and nucleus is displaced
C. It carries bad prognosis
D. Not an indication for chemotherapy

5. Following are true for clinical features of carcinoma rectum *except:*
A. Can give rise to tenesmus
B. Can present as bloody slime
C. Can present as liver secondaries
D. Can cause closed loop obstruction

6. The ideal surgical treatment for growth at 8 cm from the anal verge is:
A. Abdominoperineal resection
B. Abdominosacral resection
C. High anterior resection
D. Total mesorectal excision

7. On-table lavage of the intestines for resection and anastomosis can be done via:
A. Enterotomy
B. Colotomy
C. Enema from rectum
D. Appendicular stump

8. Local excision of malignant rectal tumour can be done if:
A. The tumour is up to 6 cm size
B. Up to 60% of the rectal wall involvement
C. Lympahtic invasion is accepted
D. Tumour is well differentiated

9. Prolapse rectum is caused by several factors *except:*
A. Birth injuries to the nerve fibres
B. Defective collagen metabolism
C. It does not start as intussusceptions
D. Deep rectovesical pouch

10. Below the dentate line squamous epithelium has:
A. No basal cells
B. Hair
C. Sweat glands
D. Pigment forming cells

11. Above the dentate line, lymphatic drainage goes to:
A. Para-aortic nodes
B. Superficial inguinal lymph nodes
C. Deep inguinal lymph nodes
D. Pudendal lymph nodes

12. Following are true for prolapsed piles *except:*
A. External sphincter grips the pile mass and cause gangrene
B. Thrombosis can occur
C. Portal pyaemia can be a complication
D. Requires hemorrhoidectomy

13. Which one of the precaution to be taken while applying band for haemorrhoids?
A. Bands are applied in grade 1 pile masses
B. Bands are applied in grade 4 haemorrhoids
C. Bands are applied below the dentate line
D. Bands should not be applied in patients who are taking anticoagulants

14. Anal stenosis is a complication of:
A. Stapler haemorrhoidopexy
B. Open haemorrhoidectomy
C. Too low application of the band
D. Cryosurgery

15. Following are true for injection line treatment of haemorrhoids *except:*
A. It is given perivascular
B. Given above the level of dentate line
C. It is painful
D. It is given in submucosal plane

16. Following are true for stapler haemorrhoidopexy *except*:
A. Recurrence rate is less
B. Less discomfort than open haemorrhoidectomy
C. Anal stenosis is not a complication
D. Ideal for 3rd or 4th degree haemorrhoids

17. In multiple fistula *in ano* and high fistula, which one of the following should not be done?
A. Biopsy of the track
B. Colostomy
C. Fistulogram
D. Multiple fistulotomy

18. Majority of the cases of fissure *in ano* are:
A. Anterior
B. Posterior
C. Anterolateral
D. Posterolateral

19. In lateral sphincterotomy:
A. Pecten fibres are ruptured
B. It is blunt sphincterotomy
C. External sphincter is divided
D. Internal sphincter is divided

20. In cases of pilonidal sinus:
A. Hair is demonstrated in the wall
B. It is congenital
C. It is known for recurrence
D. It undergoes malignant change

ANSWERS

1 C	2 D	3 C	4 D	5 D	6 D	7 D	8 D	9 C	10 D
11 A	12 A	13 D	14 B	15 C	16 B	17 B	18 D	19 D	20 C

32

Lower Gastrointestinal Bleeding

- Causes
- Clinical examination
- Investigations
- Exploratory laparotomy
- Haemobilia
- Angiodysplasia
- What is new?/Recent advances

Introduction

Lower gastrointestinal (GI) bleeding refers to bleeding which occurs beyond the ligament of Treitz. Bleeding per rectum may be a manifestation of upper GI bleeding, the causes of which have been discussed under haematemesis. In this chapter, bleeding per rectum due to lower GI causes will be discussed.

Lower gastrointestinal (GI) haemorrhage accounts for 1% of acute hospital admissions each year. Severe bleeding is that which continues for 24 hours after hospital admission or that which recurs 24 hours after resolution.

PEARLS OF WISDOM

In all, so-called the lower GI bleeding—rule out 3 important causes of upper GI bleeding, namely—oesophageal variceal bleeding due to portal hypertension, peptic ulcers—mainly duodenal ulcer bleeds and acute erosive gastritis—acute gastric mucosal lesions (AGML).

Definition

- Haematochezia: Bloody stools (LGIB or rapid UGIB)
- Melaena: Black tarry stools from digested blood. Bleeding is there for more than 8 hours.
- Massive GI tract bleeding: The bleeding which requires more than 3 units of blood transfusions in 24 hours.
- **OBSCURE:** Bleeding which persists or recurs after initial evaluation has failed (with EGD and colonoscopy). Two types:
  A. Obscure occult: Iron deficiency anaemia, faecal occult blood positive, no visible bleeding. More than 80% resolve with **no treatment.**
  B. Obscure overt: Recurrent and visible bleeding, e.g. angiodysplasia.

  Investigating a case of lower GI bleeding is like investigating a 'crime' by CBI officer. One should not jump to conclusions as soon as one cause of bleeding is found. There are innumerable examples of 'piles' being treated for bleeding, totally missing a growth above in the rectum (Table 32.1 and Key Boxes 32.1 to 32.3).

CAUSES

Depending on aetiology

KEY BOX 32.1

COMMON CAUSES OF LOWER GI BLEEDING

- Most originate in the colon or rectum—haemorrhoids, polyps, carcinoma, inflammatory bowel diseases are common causes.
- 10% from upper intestinal tract
- Small intestinal haemorrhage is usually due to arteriovenous malformations (angiodysplasia), accounting for 70–80%.
- Jejunal diverticula, Meckel's diverticula, neoplasia, Crohn's disease, and aorto-enteric fistula following a previous aortic graft are other causes of bleeding from small intestines.

I. Congenital

- Polyps: Congenital polyp, Peutz-Jeghers syndrome, Familial polyposis coli (FPC)
- Meckel's diverticulum
- Hereditary haemorrhagic telangiectasia (HHT)

PEARLS OF WISDOM

HHT is the most important inherited anomaly which produces bleeding.

II. Inflammatory
- Tubercular ulcers
- Enteric ulcers
- Crohn's ileocolitis
- Ulcerative colitis
- Necrotising enterocolitis
- **Dysentery**—amoebic, bacillary, strongyloides infestation

III. Neoplastic
- Papilloma of rectum
- Carcinoma colon, rectum
- GIST (*see* page 501, 691)
- Lymphoma
- Carcinoma small bowel

IV. Vascular
- Angiodysplasia
- Ischaemic colitis
- Vasculitis—polyarteritis nodosa
- Haemangioma

V. Clotting disorders
- Haemophilia
- Thrombocytopaenia
- Leukaemia
- Warfarin therapy
- Disseminated intravascular coagulopathy

VI. Miscellaneous
- Piles, anal fissure
- Prolapse
- Injury to the rectum
- Diverticular disease

DEPENDING ON SITE OF BLEEDING

I. Small intestine
- Peutz-Jeghers polyps
- Meckel's diverticulum
- Tubercular ulcers
- Crohn's ulcers
- Leiomyoma

KEY BOX 32.2

LOWER GI BLEEDING—TYPES

Depending upon the source:
- Small bowel bleed—5%
- Colonic bleed—95%

Depending upon the clinical manifestation:
- *Melaena*: Passage of black tarry stools (altered blood) due to slow bleeding or more proximal source of bleed.
- *Haematochezia:* Passage of bright red stools with or without clots.

KEY BOX 32.3

BLEEDING PER RECTUM WITH ACUTE ABDOMEN
- Mesenteric ischaemia
- Intussusception
- Ischaemic colitis
- Necrotising enterocolitis

II. Large bowel
- Angiodysplasia right colon
- Carcinoma colon
- Ulcerative colitis
- Dysentery
- Diverticular disease

III. Anorectal conditions
- Piles
- Prolapse rectum
- Fissure *in ano*
- Fistula *in ano* (rare)
- Injuries to the rectum

Most of the causes have been discussed in the respective chapters.

CLINICAL EXAMINATION

1. Age of the patient

- **Children and young boys:** Polyps, Meckel's diverticulum, necrotising enterocolitis.

Table 32.1 Differential diagnosis of lower GI bleeding (US statistics)

Colonic bleeding (90–95%)	Small bowel bleeding (5–10%)
Diverticular disease 30–40%	Angiodysplasia
Ischaemia 5–10%	Erosions or ulcers secondary to NSAID
Anorectal disease 5–15%	Crohn's disease
Neoplasia 5–10%	Radiation enteritis
Infectious colitis 3–8%	Meckel's diverticulum
Inflammatory bowel disease 3–4%	Neoplasia (adenocarcinoma, lymphoma)
Angiodysplasia 3%	

- **Young age group:** Piles, tuberculosis, Crohn's, dysentery
- **Middle and old age:** Carcinoma, piles, prolapse, diverticular disease.

2. Colour of blood

- **Bright red:** Piles, fissure, polyp
- **Altered blood:** Carcinoma, tubercular ulcer, Crohn's colitis, dysentery.
- **Maroon colour:** Meckel's diverticulum

3. Blood with mucus

- Intussusception
- Dysentery
- Inflammatory bowel diseases
- Carcinoma

4. Other special features

- Severe pain with bleeding: Anal fissure
- Splash in the pan: Piles
- Red currant jelly stools: Intussusception
- Streaks of blood: Anal fissure
- Bloody slime: Carcinoma rectum
- Blood with cherry-red mass coming out (piles, polyps).

5. Palpable mass abdomen

- Hard mass in the colon: Carcinoma colon
- Firm to hard mass in the right iliac fossa: Ileocaecal tuberculosis.
- Contracting mass: Intussusception

6. Rectal examination (Fig. 32.1)

- Very painful: Anal fissure
- Pedunculated mass: Rectal polyp (juvenile polyps)
- Ulcerations in the rectum: Solitary rectal ulcer
- Indurated ulcer or growth: Carcinoma rectum (Fig. 32.2).

7. Evidence of bleeding tendencies

- Purpuric spots
- Haematoma

INVESTIGATIONS

1. Proctoscopy (Fig. 32.3)

- Cherry red to pink mucosal bulges: Haemorrhoids
- Bleeding ulcer or a growth: Cancer of rectum
- Single anterior ulcer: Solitary ulcer rectum.

2. Sigmoidoscopy (Fig. 32.4)

- Multiple small pinpoint ulcers: Ulcerative colitis
- Large deep flask-shaped ulcer: Amoebic ulcers
- Multiple small polyps: Hereditary polyposis coli.

3. Colonoscopy

- It is the gold standard investigation for lower GI bleeding. It can detect 3 important diseases: Carcinoma, inflammatory bowel diseases (IBDs) and diverticular diseases (Figs 32.5 to 32.19). It can also detect ischaemic colitis, polyps and angiodysplasia. It needs to be repeated. In massive bleeding it can really tax an expert colonoscopist also.
- Colonoscopic adrenaline injections, snaring and coagulation (Argon plasma coagulation) are therapeutic advantages.

Fig. 32.1: Rectal examination

Fig. 32.2: Glove streaked with blood—carcinoma rectum

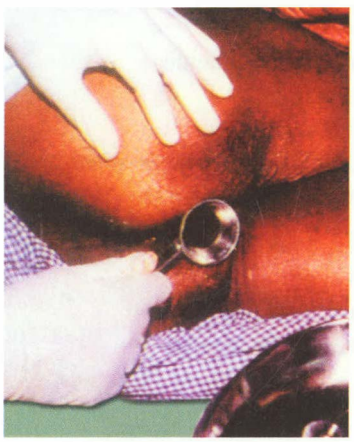


Fig. 32.3: Proctoscopy

Fig. 32.4: Sigmoidoscopy

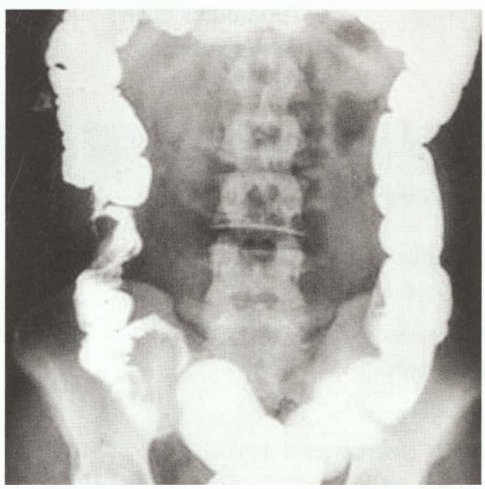


Fig. 32.5: Barium enema—carcinoma ascending colon

DIFFERENTIAL DIAGNOSIS OF LOWER GI BLEEDING (Figs 32.6 to 32.19)

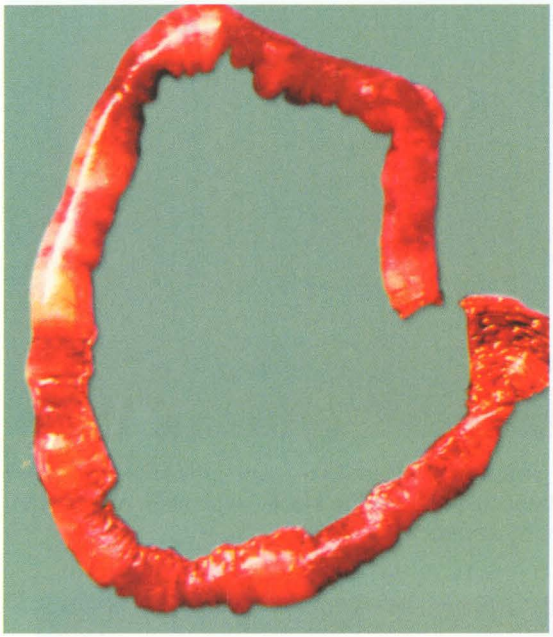


Fig. 32.6: Necrotising enterocolitis

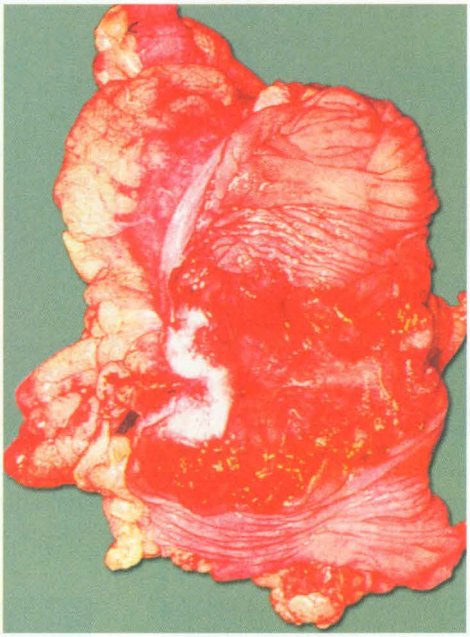


Fig. 32.7: Carcinoma colon

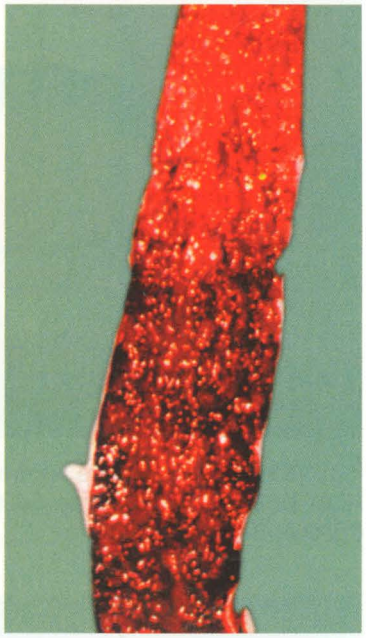


Fig. 32.8: Ulcerative colitis

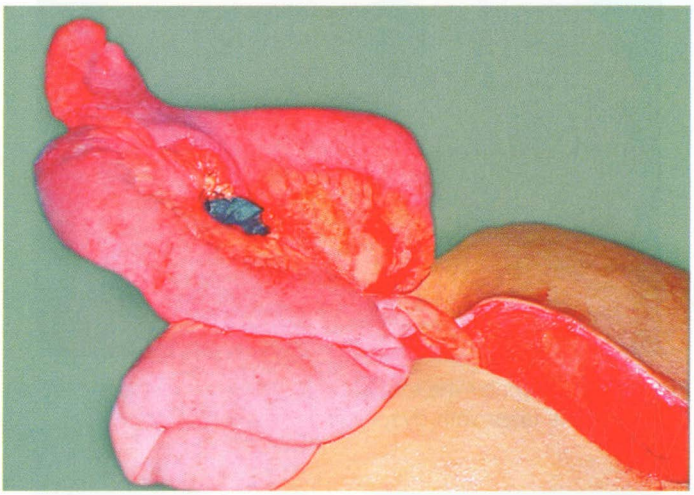


Fig. 32.9: Meckel's diverticulum

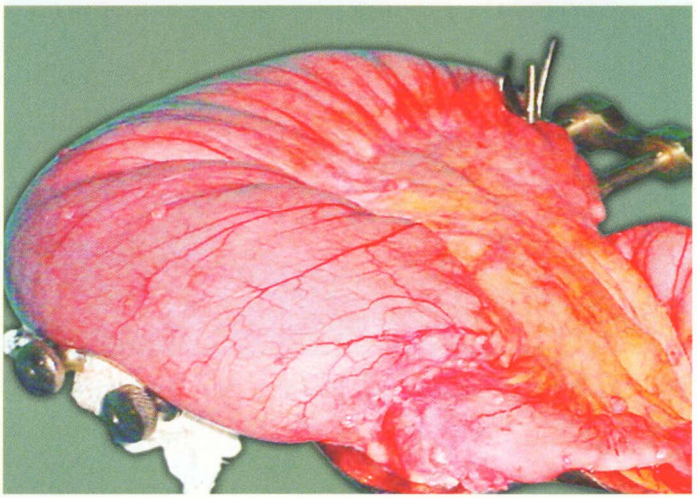


Fig. 32.10: Intestinal tuberculosis

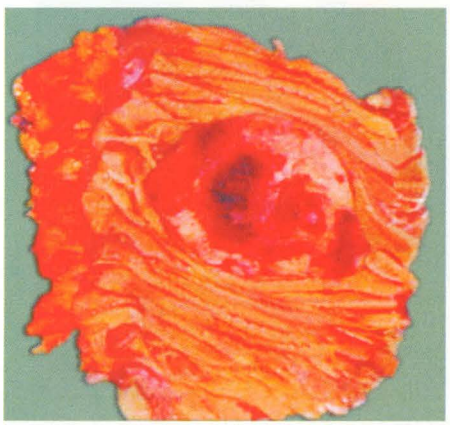


Fig. 32.11: Adenocarcinoma jejunum

Fig. 32.12: Haemorrhoids

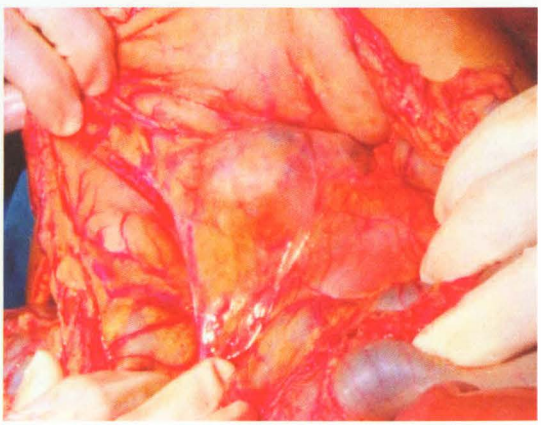


Fig. 32.13: Pancreatic pseudoaneurysm

Fig. 32.14: Peutz-Jeghers syndrome (*Courtesy:* Dr Sreevatsa, HOD, Dr Bharathi, Department of Surgery, MS Ramaiah Medical College, Bangalore)

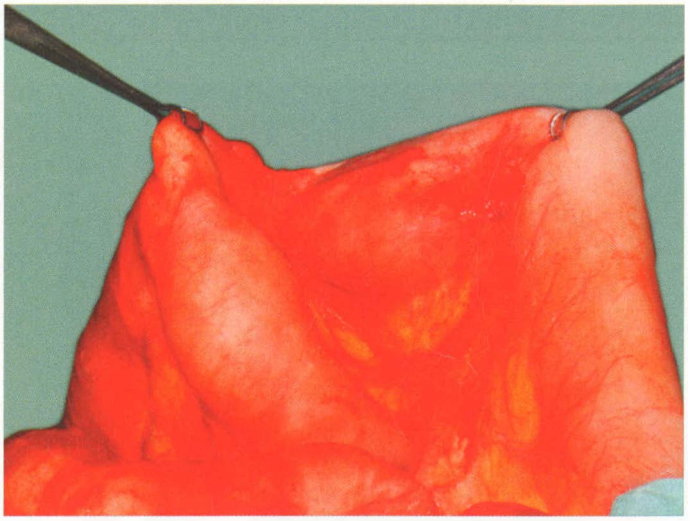


Fig. 32.15: Jejunal diverticulum from mesenteric border presented with occult blood in the stool—evaluation of anaemia, diagnosis was by enteroclysis

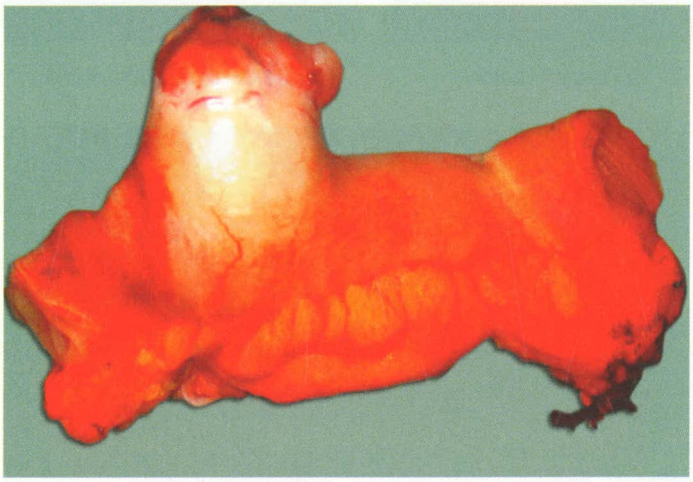


Fig. 32.16: Massive lower GI bleeding due to leiomyoma of jejunum—resected specimen

Fig. 32.17: Vascular malformation of rectum—misdiagnosed as solitary rectal ulcer with dysplasia

Fig. 32.18: Resected specimen of Crohn's disease

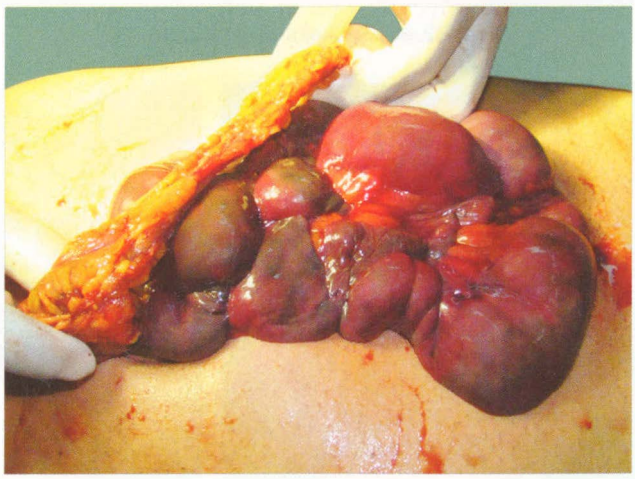


Fig. 32.19: Massive gangrene due to superior mesenteric arterial thrombosis

4. Stool examination

- Amoebiasis, bacillary dysentery
- Hookworm infestations.

5. Barium enema (Fig. 32.5)

- Irregular filling defect in the colon: Cancer colon
- Contracted pipe-stem colon: Ulcerative colitis
- Pincer ending: Intussusception
- Saw-tooth appearance: Diverticular disease.

6. Small bowel enema (enteroclysis)

- Diverticulum in the terminal ileum is Meckel's diverticulum. Multiple ulcers and stricture terminal ileum can be due to tuberculous ulcer.
- Barium studies have a little value in the presence of acute haemorrhage. They can be used in intermittent or chronic bleeding wherein endoscopy has failed to detect the cause.

7. Special investigations

They are indicated when the diagnosis of lower GI bleeding cannot be made out. They are more useful where there is active bleeding or obscure bleeding.

A. Radionuclear scanning (Fig. 32.20)

- ^{99m}Tc-labelled sulphur colloid or autologous red cells with ^{99m}Tc may be given which can detect the bleeding site. ***It is extremely sensitive, can detect as little as 0.1 ml/min of bleeding.***
- Less precise but less invasive with least complications. If ^{99m}Tc-tagged RBC scan is positive, then angiogram is used to localise the bleeding site.

B. Visceral angiography (Figs 32.20 and 32.21)

- All three vessels—coeliac, superior mesenteric and inferior mesenteric arteries are used.
- Extravasation of contrast into the bowel lumen is suggestive of a 'lesion'.
- Bleeding rate should be at least 0.5 ml/minute.
- Thus, Meckel's diverticulum, angiodysplasia, small bowel tumours, vasculitis, etc. can be diagnosed.

C. Capsule endoscopy

Definition

- It is an investigation wherein a small camera pill is swallowed to study the entire GI tract, in particular, small intestines
- This **'camera pill'** that is swallowed is disposable
- It weighs 4 grams and is 26 mm × 11 mm in size
- Parts: Video camera, lens, colour camera chip, 6 light emitting diodes. As it passes through the entire gastrointestinal tract, images are taken.
- Capsule endoscopy is useful to detect (observe) small intestinal bleeds that are missed by routine upper GI scopy and colonoscopy. The procedure takes very long time for detection of the lesions.

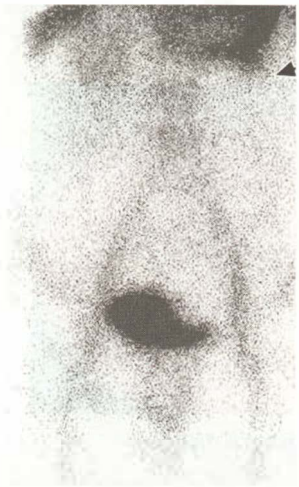


Fig. 32.20: Angiodysplasia of proximal jejunum

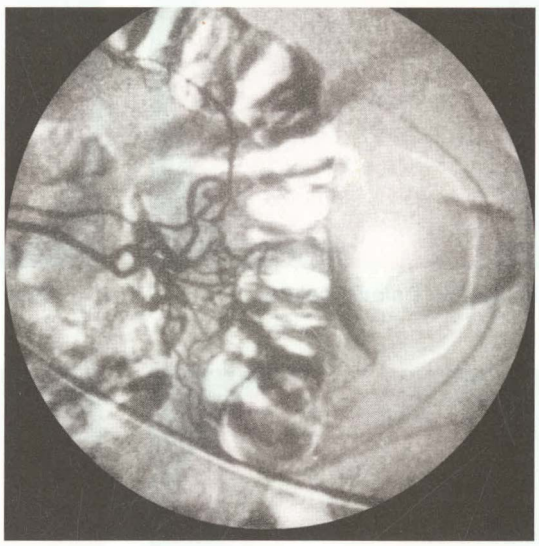


Fig. 32.21: Inferior mesenteric angiography showing leakage of dye into the lumen of sigmoid colon—sigmoid angiodysplasia

- Such bleeds are called **'obscure'** bleeds. These are very difficult problems to treat because they tend to recur.

Procedure

- Patient should be fasting overnight.
- Patient swallows the pill.
- Capsule camera sends signals and pictures—2 pictures/second are taken.
- Capsule gets deactivated in 8 hours and is passed out in stools.
- The receiver tied over patient's waist receives signals and 'endo' pictures. This is connected to computer software and pictures are obtained.

Drawbacks

- Biopsy of the lesion cannot be taken.
- It cannot detect motility disorders which are very important in GIT.
- Costly, not available in many centres.
- **Capsule** retention **can occur in 5% of cases**.

ROLE OF COLONOSCOPY/ENTEROSCOPY

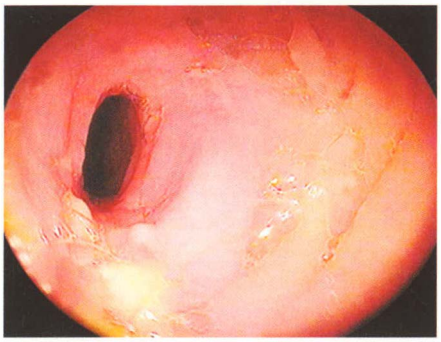


Fig. 32.22: Crohn's disease

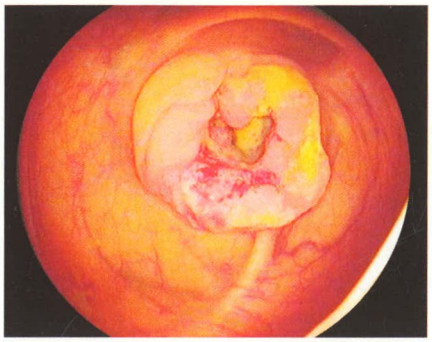


Fig. 32.23: Carcinoma caecum

Fig. 32.24: Carcinoma sigmoid colon

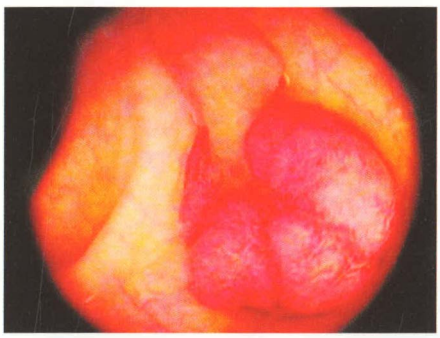


Fig. 32.25: Colonic polyps

Fig. 32.26: Colonic polyposis

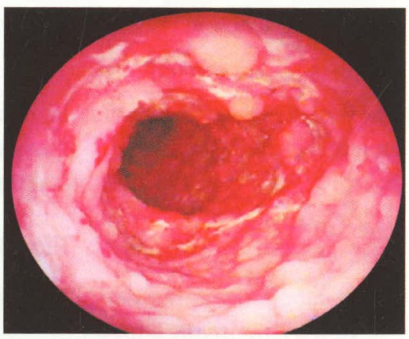


Fig. 32.27: Intestinal tuberculosis

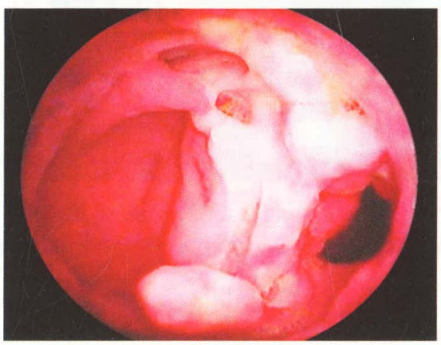


Fig. 32.28: Tuberculosis of the colon

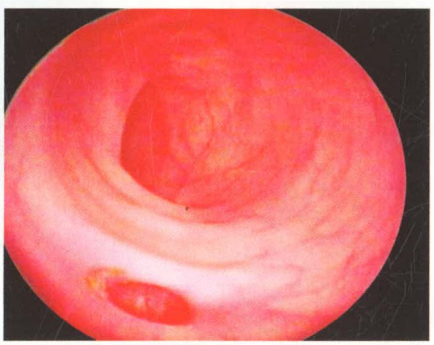
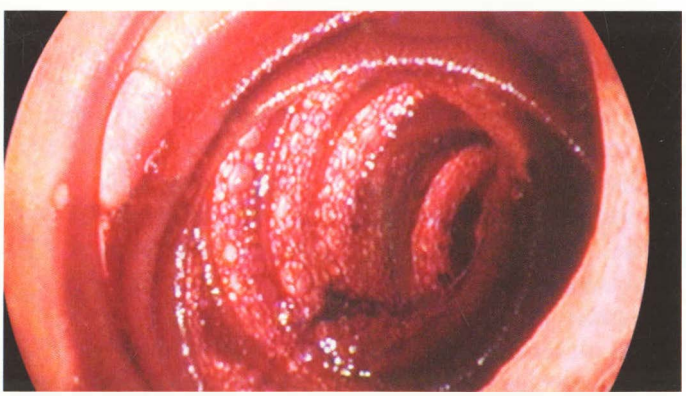


Fig. 32.29: Colonic diverticula

Fig. 32.30: Duodenal ulcer



Figs 32.31 and 32.32: Enteroscopy done at laparotomy for suspected case of angiodysplasia of the jejunum—resected successfully (*Courtesy:* For all the endoscopic pictures: Dr Filipe Alvares, Gastroenterologist, ex-KMC, Manipal)

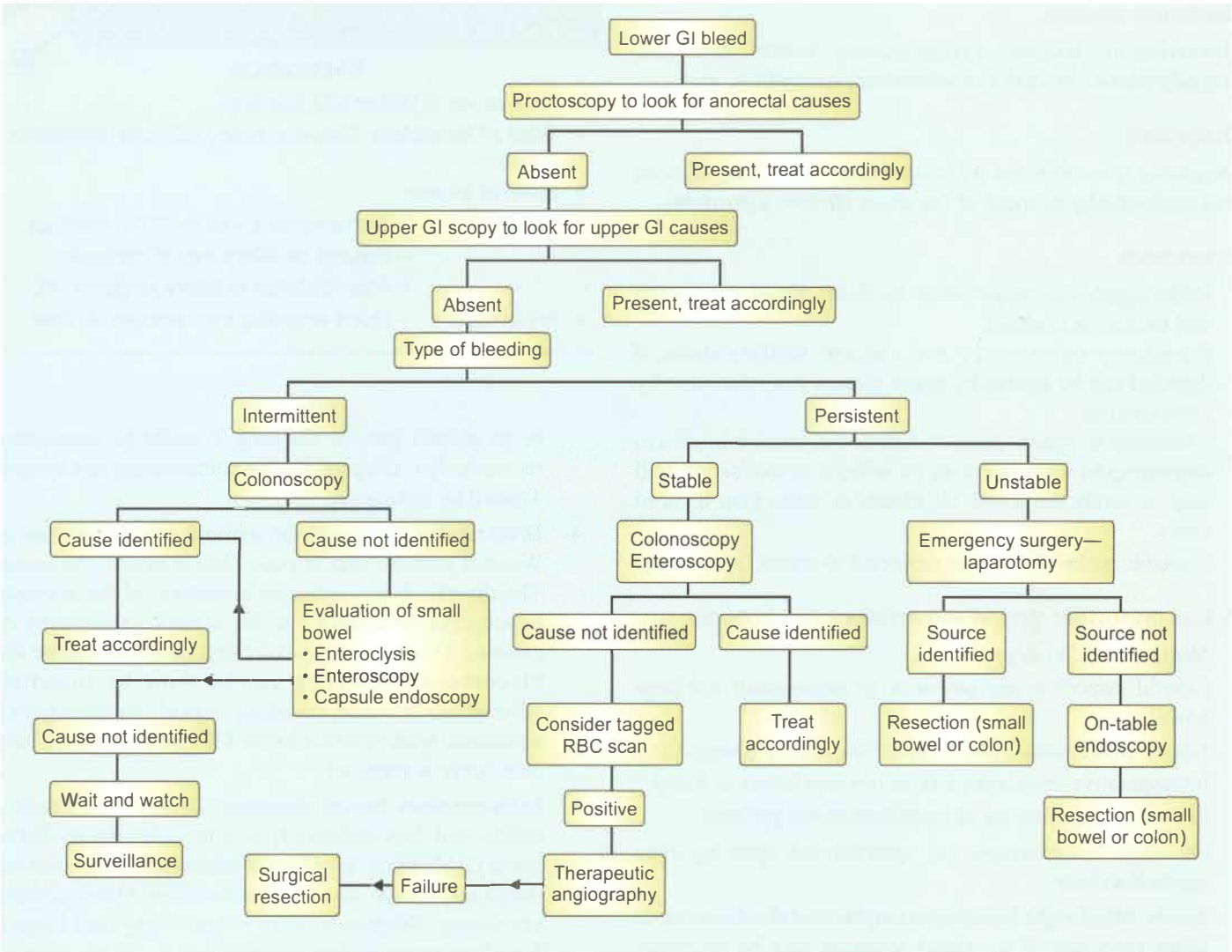


Fig. 32.33: Schematic representation of approach to lower GI bleeding
(*Courtesy:* Dr Prasad S, Associate Professor, KMC, Manipal)

Conclusion

- Thus it can detect polyps, inflammatory bowel disease (Crohn's disease), ulcers and tumours of the small intestine.
- Capsule endoscopy is an excellent tool in the patient who is haemodynamically stable but continues to bleed. Reported success rates as high as 90% in identifying small bowel pathology. It is usually well tolerated, although it is contraindicated in patients with obstruction or a motility disorder.

D. Push enteroscopy (*see* Figs 32.22 to 32.32)

- It employs 400 cm scope which is 'pushed' (hence push enteroscope) through duodenum, through DJ flexure into intestines. It needs lots of skill and expertise. It can detect leiomyoma of jejunum, small intestinal diverticular bleed causing chronic anaemia, etc.
- Extended enteroscopy is the same as enteroscopy but may take 6–8 hours till the scope travels distally with peristalsis. Up to 70% small intestine can be visualised.

PEARLS OF WISDOM

A tagged red blood cell scan can detect bleeds as low as 0.1 ml/minute but does not provide anatomical details.

Angiogram can detect bleed at 0.5 ml/min. Embolisation with metal coils can be done to correct bleed. Methylene blue can be injected to stain target segment of intestine.

MASSIVE LOWER GI BLEEDING

- Massive lower GI bleeding is defined as haemorrhage distal to the ligament of Treitz that requires more than 3 units of blood in 24 hours (Fig. 32.33).

Common causes

Diverticular disease, inflammatory bowel diseases, angiodysplasia, Meckel's diverticulum, haemobilia, etc.

Diagnosis

Diagnosis is established by colonoscopy, RBC tagged scan and angiography in most of the cases (follow algorithm).

Treatment

- Initial aggressive resuscitation by fluids, blood transfusion and treatment of shock.
- Emergency colonoscopy and vascular malformations if detected can be treated by argon plasma coagulation or by cauterisation.
- Therapeutic ***vasopressin*** infusion 0.2 units/minute *via* ***angiographic catheter*** with or without embolisation will stop or arrest the lower GI bleeds in more than 85% of cases.
- Unstable patient should be subjected to urgent laparotomy.

A few important tips at exploratory laparotomy

- Midline incision is **preferred**.
- Careful inspection and palpation of entire small and large bowel.
- Empty small bowel. Then palpate for hidden lesions.
- Intraoperative enteroscopy if no obvious lesion is found.
- Endoscopic evaluation of transilluminated gut wall.
- On table colonoscopy *via* appendiceal opening after appendicectomy.
- Rarely, **blind right hemicolectomy/subtotal colectomy or blind resection of proximal jejunum** may be necessary in obscure bleeding (keeping in mind angiodysplasia).

KEY BOX 32.4

HAEMOBILIA

- Rare cause of UGI or LGI bleeding
- **Triad of Sandblom**: Melaena, biliary colic and obstructive jaundice
- External trauma
- Iatrogenic – Transhepatic puncture (PTC, stenting)
  – Surgery on biliary tree or pancreas
  – After dilatation of biliary strictures, etc.
- Endoscopy – Blood emerging from ampulla of Vater

DIFFERENTIAL DIAGNOSIS

All the topics related to GI bleeding have been discussed in the respective chapters. Example: Haemobilia (Key Box 32.4) in page 547, haemosuccus pancreatitis in page 606, diverticular disease in page 721, inflammatory bowel diseases in page 677. Summary and important causes of the GI bleeding are given here.

I. From the colon

1. **Haemorrhoids:** These are the common causes. They cause splash in the pan. It is painless, fresh bleeding. It is one of the differential diagnoses for anaemia. Diagnosis is by proctoscopy—as cherry red spongy masses. Sigmoidoscopy is done to rule out proximal carcinoma. Treated by haemorrhoidectomy.
2. **Fissure *in ano:*** A severe painful condition of the anal canal, results in constipation, hard pellet like stools and drop of blood. Treated by lateral sphincterotomy.
3. **Carcinoma rectum/colon:** Fresh bleeding per rectum bloody slime, loss of weight, anaemia, mass abdomen in an elderly patient suggests it could be carcinoma rectum/colon. Diagnosis is by colonoscopy and biopsy. Treated by colectomy.
4. **Diverticular disease** of the sigmoid colon: Common in Western patients, diet in poor fibre is mostly the cause. The diverticuli are acquired herniation of the mucosa, hence thin. Bleeding can be occult/intermittent or massive. Diagnosis is by colonoscopy—to visualise the bleeders, endotherapy can be done by injecting adrenaline into the bleeding vessel. In emergency situations, with massive lower GI bleeding, emergency colectomy is required.
5. **Inflammatory bowel diseases:** Commonly ulcerative colitis and less commonly Crohn's disease produces lower GI bleeding. Bleeding is intermittent with mucous diarrhoea, weight loss and malnutrition. Often patients are young. Diagnosis is by colonoscopy and biopsy. Initial treatment is always conservative—salazopyrines, steroids, etc. In massive bleeding to save the life—emergency total colectomy with or without pouch may be required. In Crohn's disease, the aim is always to conserve the segment of the intestine. Resection is required only if massive bleeding is present. This is rare in Crohn's disease.
6. **Angiodysplasia** (Key Box 32.5): Vascular ectasia also called angioma, haemangiomas and arteriovenous malformations are collectively grouped under angiodysplasia. Commonly right side colon, i.e. caecum and ascending colon are affected. In the small intestines, jejunum is the most common site. Typically elderly patients present with intermittent bleeding is cause of anaemia. Usual causes of lower GI bleeding are ruled out by colonoscopy and other investigations. Suspect angiodysplasia. A few cases present with massive bleeding—a difficult problem to treat. Repeat colonoscopy, capsule endoscopy, angiography, on-table enteroscopy are the taxing investigations—all may provide no results-emergency colectomy or intestinal resection of the suspicious segment may be required.

KEY BOX 32.5

ANGIODYSPLASIA

- They are acquired lesions, seen in elderly patients
- Less rapid, but recurrent
- Caecum and right colon are common sites—Caecum is the most common site
- Small bowel (proximal) is the second common site
- Small red mucosal lesions between 2 and 10 mm, flat or raised lesions—dilated tortuous submucosal veins
- Recurrent painless and self-limiting bleeding often associated with aortic stenosis—Heyde's syndrome
- Colonoscopy is the investigation of choice
- They can be treated endoscopically—coagulation with heat probe, bipolar electrode or laser, etc. but recurrence or failure can occur
- Surgery by resecting the segment is a definitive procedure
- Angiography is rarely positive
- Enteroscopy, capsule endoscopy or intraoperative endoscopy are useful investigations
- These lesions are seen in acute renal failure, von Willebrand's disease, HHT
- Hormone treatment
- Endoscopy directed resection

7. **Ischaemic colitis:** Elderly, hypertensive patients present with diffuse abdominal pain, severe in nature, with blood in stools—it is often massive, sometimes moderate. On examination tenderness may be present on the left side of the colon. Plain X-ray abdomen supine will show **thumb printing sign** due to mucosal oedema and submucosal haemorrhage. CT scan-colonic wall thickening with posterior fat shadowing. Colonoscopy may reveal ulcers or a few changes in the splenic flexure region. If conservative measures fail such as blood transfusion, segmental colectomy may be required.
8. **Dysentery:** Various dysenteries such as amoebic, bacillary, Shigella, HIV related—all produce ulcerations in the colon resulting in blood and mucus in the stools. Gripping pain, acute in nature with or without fever and tenderness over the colon—in the right iliac fossa and in the left iliac fossa are suggestive. Diagnosis is by stool examination and colonoscopy. Treated with antiamoebic drugs or antibiotics.
9. **Radiation proctocolitis:** Usually occurs with pelvic radiotherapy, example—radiation given to treat carcinoma cervix. Most common site is rectum. Tenesmus, mucus and blood in stools common. Proctoscopy reveals ulceration. Treated with stool softening agents, 5 ASA (amino salicylic acid) topical or steroid enema.
10. **Adenoma, polyps, familial polyposis coli:** They are common in the colon. All are precursors for carcinoma colon. Often patients are young with lower gastrointestinal bleeding. Diagnosis is by colonoscopy and biopsy. Villous adenomas, polyps can be snared or excised. Always histological examination is a must.

II. From the small intestines

1. **Tubercular ulcers:** They are never massive bleeders. Patients are between 20 and 40 years old with blood and mucus in the stools, loss of weight, crampy abdominal pain, evening rise of temperature with or without pulmonary tuberculosis. On examination mass may be palpable if caecum is also involved (ileocaecal tuberculosis). Visible step ladder peristalsis indicates obstruction from a tubercular stricture or obstruction due to mass. Colonoscopy with visualisation of the terminal ileum and biopsy is the key to the diagnosis. Obstructed cases can be treated with stricturoplasty in a single stricture or resection in appropriate cases. Cases without obstruction are treated with antitubercular treatment.
2. **Crohn's ulcers:** Ileum is the commonest site—rest of the bowel can also be affected. Transmural inflammation, multiple ulcers, skip lesions are other features. Diagnosis is by CT scan, push enteroscopy and biopsy. Treatment is as for ulcerative colitis (for more details *see* page 685)
3. **Enteric ulcers:** High grade fever—enteric fever patient who has bleeding after 15 days of fever may be having enteric ulceration of the Peyer's patches with bleeding. In majority of the cases, bleeding is occult and usually stops once the disease is treated, rarely exploration and resection of the segment may be required in cases of massive bleeding cases.
4. **Meckel's diverticulum:** Children or young patients, often bleeds are intermittent, maroon coloured with or without abdominal pain. Peptic ulceration in the ectopic mucosal site in the Meckel's diverticulum causes bleeding. Colonoscopy is normal. RBC tagged technetium scan is the investigation of choice. It can pick up bleeding as little as 0.1 ml/min of bleeding. Exploration and excision of the Meckel's diverticulum is the treatment of choice.
5. **Angiodysplasias:** Small intestines are the most common sites of angiodysplasia. They are the differential diagnosis for obscure bleeds. Angiogram and small bowel push enteroscopy, capsule endoscopy are the investigations. Diagnosis is by exclusion.
6. **Small bowel tumours:** They are uncommon causes of lower GI bleeding. But they have to be kept in mind when the common causes described above are ruled out one by one. Adenocarcinoma, lymphoma and stromal tumours (GIST—gastrointestinal stromal tumours) are the few examples. GIST can affect small intestine. The mucosal ulcerations cause GI blood loss. Bleeding is not massive—can be intermittent and result in anaemia. Palpable mass sometimes massive which is bosselated, anaemia and bleeding are the triad of GIST. CT scan is the investigation of choice. Resection almost cures the

disease. Degree of malignancy is decided by the mitotic figures in pathology. Imatinib is the drug used in recurrent cases of GIST or GIST with metastasis. Patients with liver metastasis will live beyond 5–10 years with imatinib.

MISCELLANEOUS

Ischaemic colitis

- Ischaemic colitis is a non-inflammatory condition affecting splenic flexure region resulting in ischaemia and lower GI track bleeding
- Elderly hypertensive patients are commonly affected
- Often they are males
- The splenic flexure region can have relative vascularity. The exact point is called **Griffith's point** (Fig. 32.34).
- It is defined as the site of (a) communication of the ascending left colic artery with the marginal artery of Drummond, and (b) anastomotic bridging between the right and left terminal branches of the ascending left colic artery at the splenic flexure of the colon.

CLINICAL NOTES

A 28-year-old male patient had an urgency to pass stools early morning. He collapsed while passing stools, with a massive bleeding. He was brought to the hospital in a state of shock. He was resuscitated and blood transfusions were given. All investigations were normal. He had another bout of massive bleeding the next day, during which time, even an angiography could not detect the cause. Urgent laparotomy was done. A 4 cm small bowel tumour (haemorrhagic) was excised from jejunum and histology confirmed it as leiomyoma. ***Leiomyoma is called bleeding tumour of the small bowel.*** *The case history highlights the importance of* ***exploratory laparotomy. Leiomyomas are included under GIST.***

- Anastomosis at Griffith's point is present in 48%, poor or tenuous in 9%, and absent in 43%.
- Thus, it is important that in cases of ligation of inferior mesenteric artery, there is a possibility of ischaemia developing in that region.
- It can also be affected in "nonocclusive" ischaemic colitis.
- Three types have been classified—called Marston's classification:
  1. Gangrenous type
  2. Stricture type
  3. Transient type

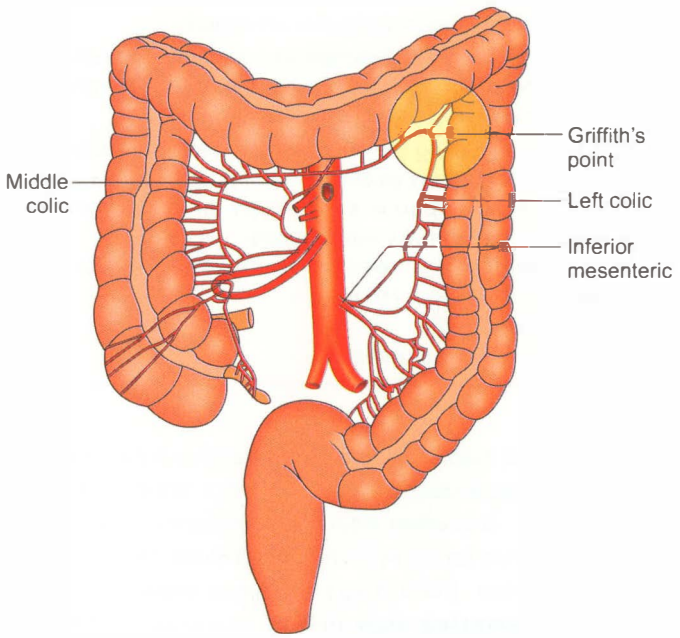


Fig. 32.34: Ischaemic colitis

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been upgraded.
- New figures, clinical notes and new investigations have been added.

MULTIPLE CHOICE QUESTIONS

1. Lower GI tract bleeding refers to bleeding:
A. Below ligament of Treitz
B. Below ampulla of Vater
C. Below Meckel's diverticulum
D. Distal to ileocaecal junction

2. Most important inherited anomaly which produces bleeding is:
A. Juvenile polyp
B. Meckel's diverticulum
C. Familial polyposis coli
D. Hereditary haemorrhagic telangiectasia

3. The ideal investigation for bleeding Meckel's diverticulum is:
A. CT scan
B. Colonoscopy
C. ^{99m}Tc-tagged RBC scan
D. Push enteroscopy

4. Following facts are true for angiodysplasia *except:*
A. They are congenital lesions
B. Right colon is the common site
C. It is one of the causes of obscure bleeding
D. Small bowel is the second common site

5. Following are true about capsule endoscopy *except:*
A. It is disposable pill
B. Ideal for small intestinal bleeds
C. Biopsy can be taken
D. Capsule retention can occur

6. Following are true for jejunal bleeding lesions *except:*
A. Carcinoma
B. Meckel's diverticulum
C. Angiodysplasia
D. Hamartomatous polyp

7. Following are true for haemobilia *except:*
A. It causes melaena
B. It causes biliary colic
C. Obstructive jaundice
D. Splenomegaly

8. Which one of the following is the cause for massive lower GI bleeding?
A. Carcinoma rectum
B. Crohn's colitis
C. Typhoid colitis
D. Diverticulitis of the colon

9. Which one of these causes bleeding with septic shock?
A. Carcinoma colon
B. Ulcerative colitis
C. Mesenteric ischaemia
D. Angiodysplasia

10. Which one of these causes painless and massive bleeding per rectum?
A. Angiodysplasia
B. Sigmoid volvulus
C. Necrotising enterocolitis
D. Mesenteric ischaemia

ANSWERS

1 A	2 D	3 D	4 A	5 C	6 B	7 D	8 D	9 C	10 A

33

The Appendix

- Development and anomalies
- Surgical anatomy
- Acute appendicitis
- Differential diagnosis
- Complications
- Appendicular mass
- Faecal fistula
- Neoplasm
- Mucocoele
- Valentino appendix
- Post-appendicectomy sepsis—case report
- What is new?/Recent advances

Introduction

Acute appendicitis is the most common emergency encountered by the general surgeons. Men have slightly increased incidence of acute appendicitis compared to women. Incidence is 11 per 10,000 persons/year. Appendicectomy is a simple surgery, no doubt, but sometimes it can be very difficult and disappointing—sometimes one may not be able to find the appendix. Hence, appendicectomy should not be taken lightly. The choice of surgery today is laparoscopic appendicectomy—one advantage being one can look into all quadrants of the abdomen—not to miss other causes such as perforated duodenal ulcer (*see* later **Valentino appendix**), etc.

Few historical events

- 1736: Claudius Amyand removed inflamed appendix from the hernia sac of a boy.
- 1886: Reginald Fitz of Boston identified the appendix as the primary cause of the right lower quadrant inflammation. He coined the word appendicitis.
- 1889: Charles McBurney suggested early laparotomy and removal of the appendix. He also describe the McBurney point of maximum tenderness.
- The first laparoscopic appendicectomy was described by Kurt Semm.
- 2009: First transvaginal removal of the appendix by Santiaggo Horgan and Mark A. Talamini—a procedure called **NOTES**—**N**atural **O**rifice **T**ransluminal, **E**ndoscopic, **S**urgery (more details on page 1178).

DEVELOPMENT AND ANOMALIES

- Embryologically, the appendix and caecum develop as outpouchings of the caudal limb of the midgut loop in the sixth week of human development. By the fifth month, the appendix elongates into its vermiform shape, hence called vermiform appendix. At birth, the appendix is located at the tip of the caecum but due to unequal elongation of the lateral wall of the caecum, the adult appendix typically originates from the posteromedial wall of the caecum, caudal to the ileocecal valve. A few anomalies are given below:

1. **Duplication of the appendix** is one anomaly which is further divided into following ways

   Type A: Single caecum—partial duplication
   Type B: Single caecum and 2 separate appendices
   Type C: Double caecum with each one having one appendix (Figs 33.1 and 33.2)
2. **Situs inversus:** In this condition appendix is found on the left side. Adds confusion in the diagnosis of acute appendicitis
3. **Subhepatic appendix:** It happens in malrotation of the gut. Patients with subhepatic appendicitis may complain of pain in the right lower quadrant. A McBurney incision is usually given only to find no appendix in that location. Laparoscopy has the advantage of looking into all quadrants of the abdomen.
4. **Congenital absence of the appendix** is rare.

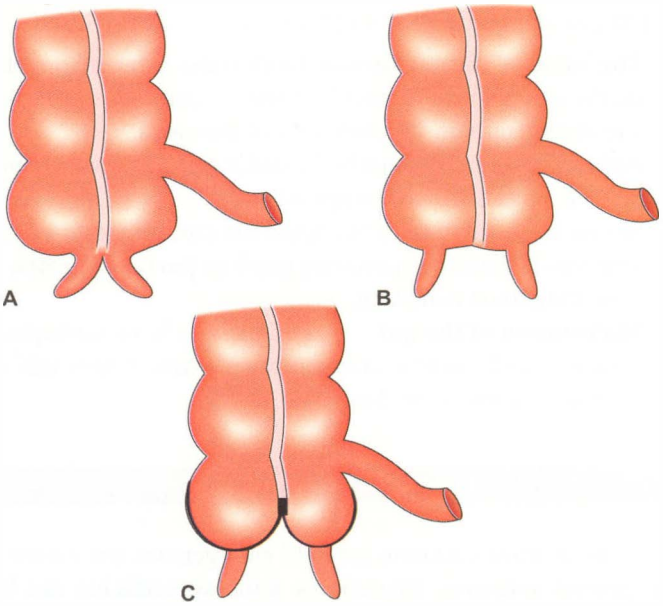


Fig. 33.1 A to C: Anomalies of the appendix (*see* text for details)

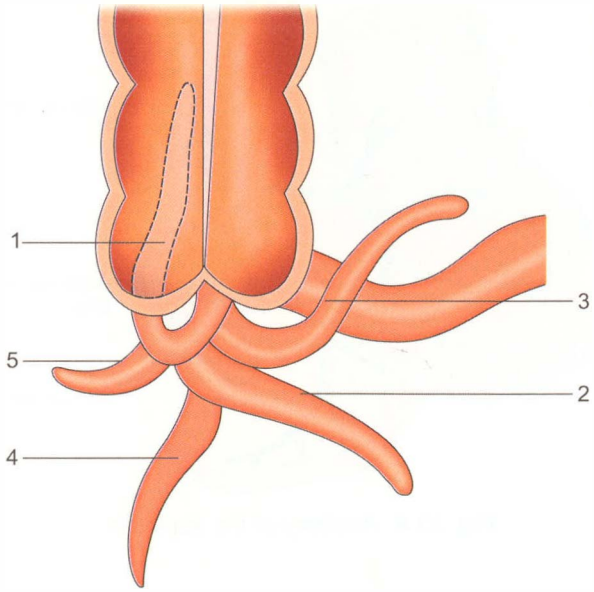


Fig. 33.3: Positions of the appendix—*see* text for numbers

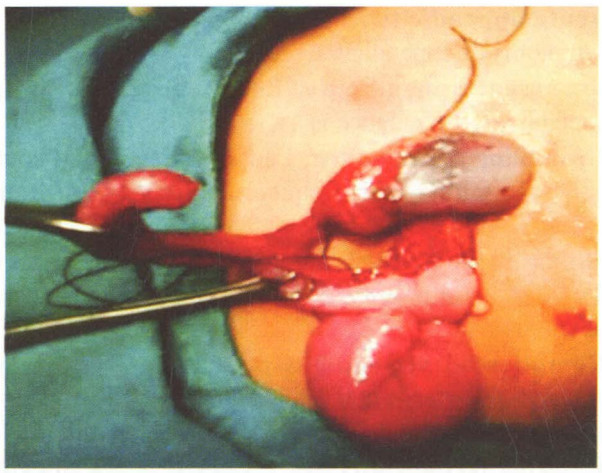


Fig. 33.2: Appendicular duplication and gangrene in one of the moieties

SURGICAL ANATOMY OF THE APPENDIX[1]

- It is 8–10 cm long, may vary from 3 to 30 cm in length.
- It is situated 2 cm posteromedial to ileocaecal junction, at the point of convergence of the three taeniae coli.
- It is the primary cause of lower abdominal pain on the right side.

Positions of the appendix (Fig. 33.3)

1. Retrocaecal in about 70% of patients (12 o'clock)
2. Pelvic in 20% of cases (4 o'clock)
3. Preileal and postileal (2 o'clock)
4. Subcaecal (6 o'clock)
5. Paracaecal
6. **Subhepatic appendix** is associated with subhepatic caecum. It occurs due to **malrotation of the gut** (this position is not depicted in the figure)

Layers of the appendix

- Mesoappendix is the continuation of mesentery of the ileum above. It comes down carrying blood vessels in the meso-appendix.
- Appendix has a serosa and a mucosa lined by columnar epithelium (similar to intestinal mucosa) between which are the circular and longitudinal muscle fibres.
- **Submucosa has rich lymphoid follicles** (lamina propria). The lymphatic tissue decreases as age advances. Hence, incidence of **appendicitis is less after the age of 30 years**.
- Appendicular orifice is occasionally guarded by an indistinct semilunar fold of mucous membrane, known as ***Valve of Gerlach***.

Blood supply of the appendix

- Appendicular artery is a branch of ileocolic artery. Accessory appendicular **artery of Sheshachalam** (a branch of posterior caecal artery) is a branch of ileocolic artery, which runs in the mesoappendix (Fig. 33.4).
- Veins follow the artery and end in the superior mesenteric vein, thus draining into portal vein. This is the reason for development of pylephlebitis in cases of suppurative appendicitis.

[1]Appendix secretes immunoglobulins particularly IgA. So it is not considered as a vestigial organ anymore. However, appendicectomy is not associated with any immunological compromise.

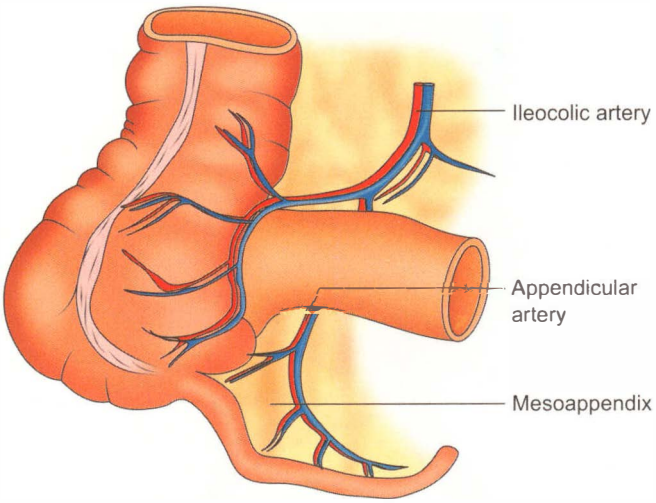


Fig. 33.4: Anatomy of the appendix

Surgical importance

- Suppurative appendicitis can give rise to pylephlebitis (inflammation of portal venous radicles).

Locating the appendix

- Trace the taenia coli or trace ileal loops at laparotomy. Taenia coli point to the base of the appendix. However, surface marking of the appendix is done as follows: Draw a line from anterior superior iliac spine to the umbilicus. The junction of lateral 1/3rd and medial 2/3rds of this line indicates the location of appendix. This is the point of maximum tenderness in appendicitis. This is called **McBurney's point** (Fig. 33.5).

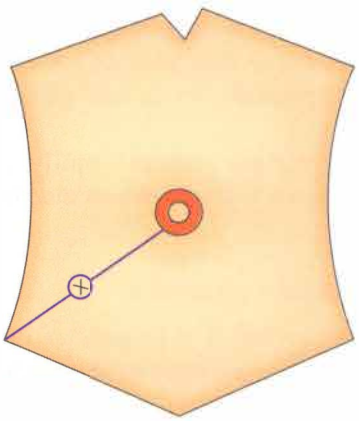


Fig. 33.5: McBurney's point

Lymphatics

- The lymphatic channels which are 4 to 6 in number drain into ileocolic nodes, ileocaecal nodes and appendicular nodes in mesoappendix.

Surgical anatomy and significance

1. **The area of the maximum tenderness** in acute appendicitis is called McBurney's, point — corresponds to the site of appendix in vast majority of the cases.
2. Appendicular artery must be ligated in open or laparoscopic method — **to free mesoappendix.**
3. Severe inflammation of the appendix can spread to portal vein via ileocolic vein and can result in **portal pyaemia,** a very dangerous condition.
4. **Malrotation of the gut** — appendix may be in subhepatic region — to be kept in mind in cases wherein appendix is not found in the right iliac fossa.

ACUTE APPENDICITIS

It is one of most common surgical emergencies encountered by general surgeons. Sometimes acute appendicitis can be dangerous (Key Box 33.1).

Definitions

- **Acute appendicitis:** Sudden appearance of signs and symptoms of appendicitis.
- **Recurrent appendicitis:** Recurrent attacks of acute appendicitis—incidence is 15 to 25%.
- **Grumbling appendicitis:** Low grade recurrent bouts of colics, vomiting with frequent admission, self-limiting cases.
- **Simple appendicitis:** If duration of symptoms is less than 48 hours or imaging does not show any abscess or phlegmon.
- **Complicated appendicitis:** Acute appendicitis with perforation or large abscess/phlegmon.
- **Pseudoappendicitis**: Acute ileitis mimics appendicitis following ***Yersinia*** **infection**. It can also be due to **Crohn's** disease.

KEY BOX 33.1

WHY APPENDICITIS IS DANGEROUS?

- The appendix is a *cul-de-sac* (closed at one end) and can be easily blocked
- The appendicular artery is an end-artery (gangrene can occur fast)
- Inflammatory oedema causes easy and early thrombosis of appendicular artery
- The appendix has thin muscular coat. Hence, perforates easily
- The lumen of the appendix is very narrow—1–3 mm in diameter
- Closed loop obstruction: Intraluminal pressure builds up as the appendicular mucosa secretes fluid resulting in mucosal ischaemia. Slowly bacterial overgrowth and translocation occurs

- **Stump appendicitis:** It is the inflammation and infection of appendicular stump, if a big stump is left behind (post-operative cases). It may require stump appendicectomy. It is important to ligate and divide at the base of the appendix to avoid this complication (more so in laparoscopic appendicectomy).

Aetiology

1. Racial and dietary factors

- It is more common in white race than in coloured persons. Young males are affected more often.
- It may be related to Westernisation of food—a diet rich in meat precipitates appendicitis and ***a diet rich in fibre (cellulose) protects the person from appendicitis.***

2. Familial susceptibility: It is related to having a ***long retrocaecal appendix*** in which case the blood supply is diminished to the distal portion and may precipitate appendicitis.

3. Socioeconomic status: Appendicitis is common in middle class and rich people. The exact reasons are not known.

4. Obstructive theory: Obstruction to the lumen of the appendix due to faeco-liths, worms, ova, cysts of *Entamoeba* causes obstructive appendicitis. It is seen only in one-third cases.

5. Nonobstructive theory: It is due to bacteria such as *E. coli,* Enterococci, Proteus, Pseudomonas, Klebsiella and anaerobes which produce diffuse inflammation of appendix and cause appendicitis. This seems to be more common cause than obstruction.

Pathology

I. In nonobstructive cases (catarrhal appendicitis)

- Process of inflammation is slow and gradual.
- A mild attack may completely resolve or mucosal and submucosal oedema can occur (Key Box 33.2).
- Ulceration of the appendix results in slow bacterial invasion of lymphoid tissue.
- Gangrene and perforation are rare.

KEY BOX 33.2

NONOBSTRUCTIVE THEORY IN ACUTE APPENDICITIS

- This is seen in two-thirds of the cases. Hence, more common than obstructive theory
- Bacterial or viral infection is the cause
- It causes mucosal ulceration
- This is followed by bacterial invasion
- The decrease in the incidence of enteric fever in the Western world has decreased incidence of acute appendicitis—a support for infective theory
- In many cases of appendicitis, the appendix is not dilated (against obstructive theory)

II. In obstructive cases

- **Symptoms are abrupt, vomiting is more, pain is more and tenderness is more.**
- It is a more dangerous variety.
- Appendix looks inflamed, with congested blood vessels. Tip especially looks more inflamed. As the inflammation is more severe, the outer aspect looks dull and purulent exudates may be seen. Areas of blackening or green colour indicates gangrene or necrosis with perforation. In acute inflammation neutrophils are dominant and in cases of gangrenous appendicitis, vascular thrombosis is a feature. The important pathological events can be summarised as follows—due to obstruction, the contents get infected fast and the tension increases. The appendix becomes a closed loop, which results in septic thrombosis of vessels. Gangrene of appendix, perforation, peritonitis, followed by a local abscess can occur (Fig. 33.6).
- In **children, greater omentum is very thin**. Hence, it cannot localise the infection. In adults, omentum is like a fatty apron which localises the infection.
- In **aged patients,** because of atherosclerosis, **gangrene** occurs **very fast** resulting in peritonitis. Obstruction is caused by faecoliths, worms and bands which cause tenting. ***Obstructed appendicitis is one of the examples for closed loop obstruction.*** Other causes are volvulus, carcinoma hepatic flexure, etc.
- Common bacteria encountered in acute appendicitis are *Bacteroides fragilis, Escherichia coli, Clostridium perfringens, Streptococcus faecalis, Pseudomonas aeruginosa*, etc.

Clinical features

The peak incidence is in the second and third decades. Very uncommon before the age of two.

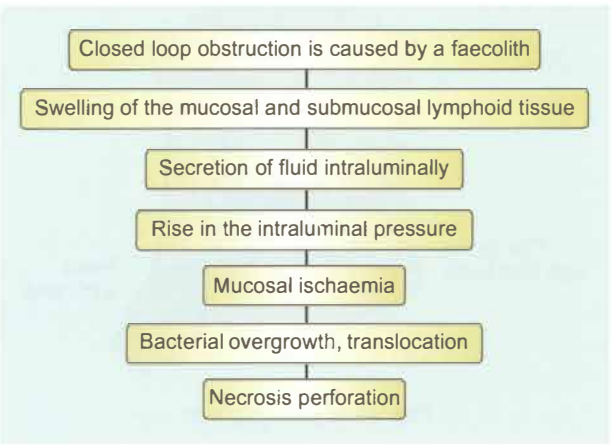


Fig. 33.6: Pathogenesis of appendicitis

Symptoms

- **Pain** is severe, colicky type, initially felt in the umbilical region and it is due to ***distension of appendix.*** This is a visceral pain. After a few hours, the pain localises to the right iliac fossa. It is a ***somatic pain*** which is due to ***inflammation of parietal peritoneum.*** This is called ***shifting pain of acute appendicitis*** (Fig. 33.7). This is called ***migratory pain—most reliable symptom of acute appendicitis.***
- Normal appendix is mobile. So, the site of maximum pain and tenderness can vary.
- **Vomiting** occurs once or twice due to reflex pylorospasm. It contains **stomach contents**. However, it is never frequent such as in intestinal obstruction.
- Appendicitis is unlikely in patients with normal appetite. Usually patients have **anorexia.**
- **Fever** is of low grade (around 100°F) and indicates bacterial inflammation.

PEARLS OF WISDOM

Pain first, followed by vomiting and then by fever is called Murphy's[1] triad of symptoms of acute appendicitis (Murphy's syndrome).

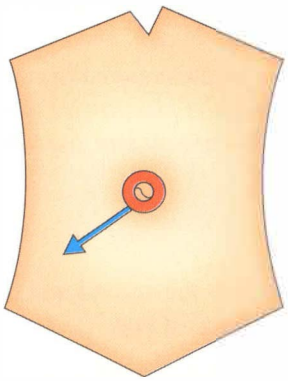


Fig. 33.7: Shifting pain (migratory pain)—most reliable symptom

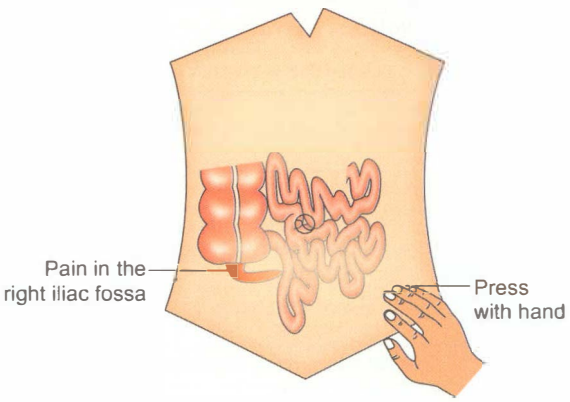


Fig. 33.9: Rovsing sign

[1]Can you find out what is Murphy's sign and Murphy's punch test?

- **Haematuria** is uncommon and it is due to inflammation of retrocaecal appendix which irritates the ureter in the retroperitoneum.
- **Constipation** is the usual feature, except in pre- and post-ileal appendicitis, where they produce diarrhoea due to irritation of ileum.

Signs

1. ***Cough tenderness*** (Fig. 33.8) indicates inflammation of parietal peritoneum. This is an important physical sign which differentiates acute appendicitis from right-sided ureteric colic.
2. ***Tenderness and rebound tenderness*** are present at McBurney's point. Rebound tenderness is called **Blumberg sign.** It is due to inflammation of the parietal peritoneum. This physical sign can be elicited in all cases of peritonitis.
3. ***Guarding and rigidity*** are present in the right iliac fossa. However, guarding and rigidity of back muscles (erector spinae) indicates retrocaecal appendicitis.
4. ***Rovsing sign:*** Palpation of left iliac region of abdomen produces pain in the right iliac region. It is because of displacement of colonic gas and small bowel coils impinging upon the inflamed appendix (Fig. 33.9).
5. ***Hyperaesthesia*** in the Sherren's triangle (Fig. 33.10): It is formed by anterior superior iliac spine, umbilicus

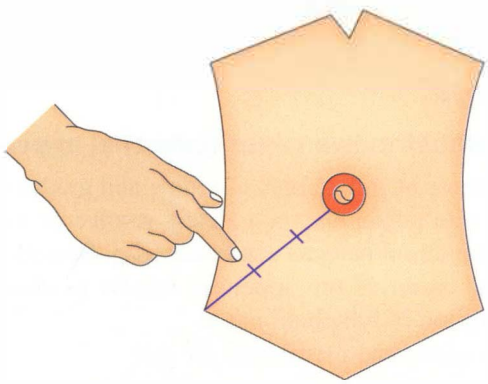


Fig. 33.8: Cough tenderness (Dunphy's sign)

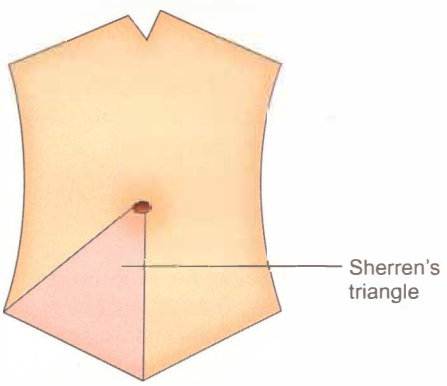


Fig. 33.10: Hyperaesthesia in the Sherren's triangle

KEY BOX 33.3

VARIATIONS IN ACUTE APPENDICITIS

1. **Retrocaecal**: Silent (no rigidity in the right iliac fossa)
2. **Pelvic:** Causes diarrhoea
3. **Postileal:** Causes diarrhoea—called missed appendix
4. **Subhepatic:** Manifests as pain in the right iliac fossa, very difficult to remove from gridiron incision
5. **In pregnancy:** The location of the pain is shifted higher up and laterally

and pubic symphysis. It is due to irritation of lower abdominal nerves.

6. ***Cope's psoas test:*** Seen in retrocaecal appendicitis. There will be irritation of psoas major which produces flexion at the hip. If any attempt is made to extend the hip, it produces pain.
7. ***Cope's obturator test:*** Seen in pelvic appendicitis due to irritation of the obturator muscle. Flexion and medial rotation produces pain.
8. ***Features of generalised peritonitis*** are seen only when there is a rupture. Gangrene and perforation is more common in elderly patients because of atherosclerosis. In infants, omentum is very thin without much of fat. Hence, diffuse peritonitis occurs very fast.
9. ***Rectal examination:*** There is tenderness in the right rectal wall—**differential tenderness**.
10. ***Per vaginal examination:*** Presence of ovarian mass, tenderness on **movement** of cervix, adnexal tenderness may suggest obstetric pathology.
    - Signs and symptoms vary depending upon the location (Key Box 33.3).

Investigations

1. **Total WBC count** is almost always increased above 10,000 cells/mm^3, in most of the patients (95%).
    - A very high white blood cell count (> 20,000/mm^3) suggests complicated appendicitis with gangrene or perforation.
2. **Urine examination** is mainly to rule out urinary tract infection, haematuria and sometimes pyluria.
3. C-reactive protein is elevated in any inflammatory condition such as appendicitis. Elevated in the first 12 hours of acute inflammation very non-specific.
4. **Plain X-ray abdomen erect** is taken to rule out perforation and intestinal obstruction. It may show dilated small bowel loops in the right iliac fossa.

PEARLS OF WISDOM

Presence of faecolith is highly suggestive of acute appendicitis in plain X-ray.

5. **Abdominal ultrasound** to rule out other causes including gynaecological causes. Ultrasound can demonstrate a non-compressible, aperistaltic tubular organ with a thick wall. It can be used to elicit probe tenderness (sensitivity of 85%, specificity 90%).

Advantages

- It is a simple bedside investigation
- Economical
- Can confirm acute appendicitis in about 50% of the patients
- Appendicolith, pericaecal fluid collection or inflammation can be diagnosed—indirect features of acute appendicitis
- More sensitive and specific in children—thin abdominal wall.

Disadvantages

- It is operator-dependent
- It is not a choice in fatty obese patient
- Gas within the dilated intestine may obscure the appendix

6. **CECT—Contrast Enhanced CT scan is the investigation of choice** (sensitivity 90%, specificity 90%), specially when diagnosis is not established or in unclassic cases. All the findings mentioned in the ultrasound can also be defined by CT scan (Fig. 33.11).

Advantages

- More objective
- Sensitivity and specificity is almost about 95%
- Helps to rule out carcinoma caecum, duodenal perforation, acute pancreatitis, etc.

Disadvantages

- Pregnant woman—it is contraindicated
- In children—better to avoid it for the fear of radiation exposure and risk of cancer developing at a later date
- Expensive, long time for the contrast to reach the site
- Low fat, sensitivity is less

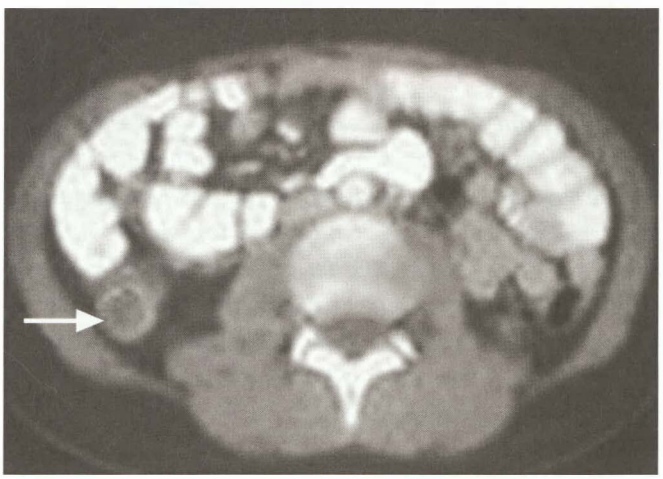


Fig. 33.11: CT scan in acute appendicitis — showing a faecolith

- Allergy to contrast and contrast nephropathy (dehydration, high creatinine, diabetics precipitating factors).

SCORING SYSTEM

To avoid negative appendicectomies, many scoring systems have been developed considering signs, symptoms and investigations. Most commonly used **Alvarado scoring system** is given in Table 33.1.

Score less than 5	:	Not sure
Score 5–6	:	Compatible
Score 6–9	:	Probable
Score more than 9	:	Confirmed

- Even though Alvarado scoring is highly suggestive of appendicitis, it is only a simple and cost-effective scoring system. This can be applied when sophisticated investigations such as ultrasonography and CT scan are not available.

Table 33.1 Alvarado scoring system

Features	Score
Symptoms:	
Migrating RIF pain	1
Anorexia	1
Nausea, vomiting	1
Signs:	
Tenderness RIF	2
Rebound tenderness	1
Elevated temperature	1
Laboratory:	
Leucocytosis	2
Shift to left	1
Total	**10**

CLINICAL NOTES

A 65-year-old lady was examined for feature of acute appendicitis of 8 hours duration. On examination, she had McBurney tenderness but a vague mass was palpable. It is unusual for an appendicular mass to appear within 8 hours following appendicitis. Ovarian pathology was considered and gynaecological opinion was requested. It was normal. CT scan was done. It revealed mucocoele of the appendix (8 cm size). She underwent lower midline laparotomy and it was removed.

CT scan gave a correct diagnosis and it guided the treatment policy.

Surgical wisdom: Symptoms, signs (tenderness in McBurney point) with increased total counts, often you do not need any imaging tests.

DIFFERENTIAL DIAGNOSIS OF ACUTE APPENDICITIS

Innumerable conditions may mimic some signs of appendicitis. A few important conditions have been considered here.

In children (Fig. 33.12A to D)

A. Enterocolitis is common in children. It presents with severe diarrhoea with blood and mucus in the stools.

B. Meckel's diverticulitis can present with abdominal pain, vomiting, fever—signs and symptoms are similar to acute appendicitis (difficult to differentiate clinically).

C. Worm ball is common in children in the developing countries. However, features of intestinal obstruction will be present.

D. Acute iliac/mesenteric lymphadenitis—non-shifting pain and rebound tenderness are absent. It is viral in origin and self-limiting. Neck nodes will give clue to the diagnosis.

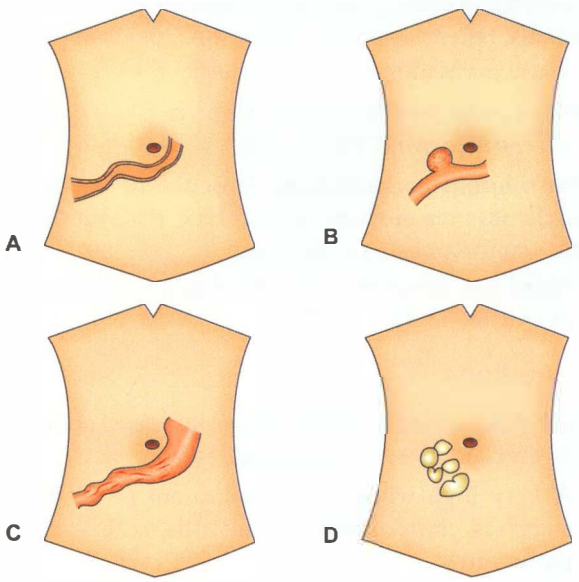


Fig. 33.12A to D: In children (*see* text)

In young adults (Fig. 33.13A to D)

A. Right-sided ureteric colic: Haematuria, severe pain from loin to groin, absence of cough tenderness help in excluding acute appendicitis.

B. Amoebic typhlitis is associated with diarrhoea, blood in the stools and tenderness in left iliac fossa (Manson Barr's amoebic point of tenderness).

C. Torsion of undescended testis: Absence of testis in the scrotum clinches the diagnosis.

D. Meckel's diverticulitis

E. Yersinia ileitis: Acute, self-limiting inflammation of the ileum caused by *Yersinia pseudotuberculosis*.

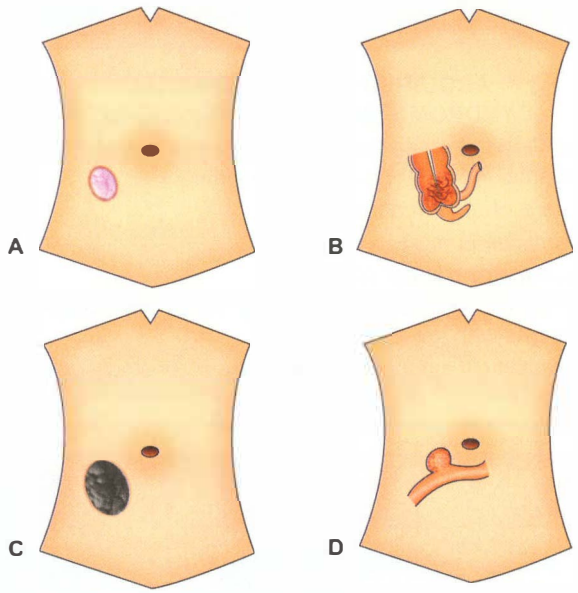


Fig. 33.13A to D: In adults (*see* text)

In middle age (Fig. 33.14A to D)

A. Acute pancreatitis: Inflammatory exudate collects and gravitates in the right iliac fossa resulting in pain, guarding and rigidity in the right iliac fossa. History of alcohol intake, severe backache and tenderness in the epigastrium helps in diagnosing acute pancreatitis.

B. Perforated duodenal ulcer can present with pain in the right side of the abdomen due to similar causes mentioned above.

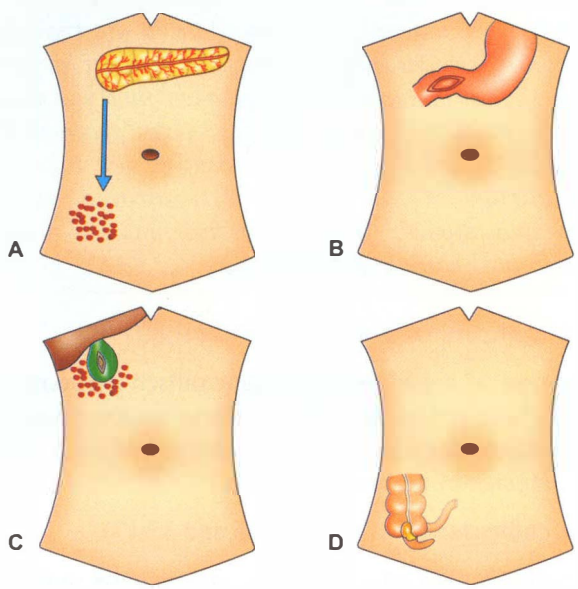


Fig. 33.14A to D: In middle age (*see* text)

C. Acute cholecystitis can also present with features of acute appendicitis. However, it is common in elderly females.

D. Pain in the right iliac fossa and tenderness is due to dilated intestinal loop peristalsis as in ileocaecal tuberculosis or carcinoma caecum. Presence of an irregular, hard mass suggest carcinoma caecum.

In females (Fig. 33.15A to D)

A. Ruptured ectopic gestation: Missed periods, features of haemorrhagic shock (pallor), extreme tenderness on movement of cervix during *per vaginal* examination clinches the diagnosis.

B. Pelvic inflammatory diseases:

- These are group of inflammatory conditions affecting young women
- Tubo-ovarian sepsis, salphingitis and endometriosis are grouped under this
- Pain is bilateral, fever is higher degree, no anorexia are a few features
- Tenderness is present on both iliac fossae on deep palpation—no cough tenderness
- Vaginal discharge helps in the diagnosis
- ***Chlamydia trachomatis*** and ***Neisseria gonorrhoeae*** culture to be done.

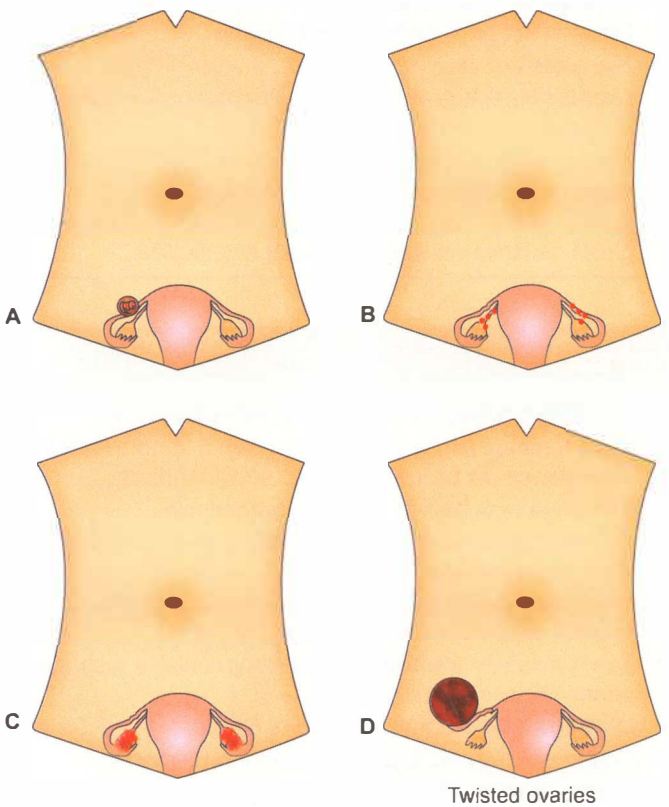


Fig. 33.15A to D: In females (*see* text)

C. **Midmenstrual** (mittelschmerz) rupture of ovarian follicle occurs about 14th to 16th day and can produce abdominal pain.

D. **Torsion of ovarian cyst** produces very severe abdominal pain with a mass.

PEARLS OF WISDOM

Any female patient with right-sided lower abdominal pain should undergo a gynaecological examination to rule out the causes mentioned above, before subjecting to appendicectomy.

Systemic diseases

1. **Pleurisy** and **pneumonia.**
2. **Porphyria:** Violent intestinal colic occurs due to spasm. It is precipitated by barbiturates. Urine is orange-coloured and when it is exposed to sunlight, the colour changes to amber.
3. **Pott's spine** causes compression of nerve roots—radicular pain.
4. **Preherpetic pain** of 10th and 11th dorsal nerve is located over the same area. Marked hyperaesthesia is present.
5. **Purpura** and bleeding disorders.

KEY BOX 33.4

CHILDREN AND ACUTE APPENDICITIS

- Appendicitis is rare under 2 years of age because lymphatic tissue is not yet developed by that time.
- Signs are not very well located.
- Greater omentum is very thin. Perforation peritonitis is common.
- Hence, early surgery is recommended.
- Open or laparoscopic method is followed.
- Remember to rule out acute mesenteric lymphadenitis (viral), Yersinia ileitis and Meckel's diverticulitis.

KEY BOX 33.5

PREGNANCY AND ACUTE APPENDICITIS

- Most common cause of abdominal pain and **nonobstetric emergency in pregnancy** is acute appendicitis.
- Incidence may be 1 to 1.5/1000 pregnancies.
- Symptoms of nausea and vomiting are confused for **morning sickness.**
- Migration of pain need not be present. Leukocytosis is seen in pregnancy cases—it is normal occurrence.
- Tenderness is **shifted** because appendix is displayed superiorly and laterally.
- **Ultrasound** is the investigation of choice.
- Treatment is by laparotomy and appendicectomy.
- **Foetal loss is 3%** but with perforation, it is 30%.
- Maternal mortality rate in perforated appendicitis is 4%.

Few special situations

One should be careful and be firm in decision making of appendicectomy in these cases (Key Boxes 33.4 to 33.6).

KEY BOX 33.6

ACQUIRED IMMUNODEFICIENCY SYNDROME (AIDS) AND APPENDICITIS

- Incidence of acute appendicitis is more common in AIDS patients—4 fold than non-AIDS patients
- Pain is chronic than acute
- Diarrhoea is more common
- Leukocytosis is not common
- Delay in the presentation may be present especially patients with low CD count
- Interestingly outcome or results are surprisingly good after surgery

CLINICAL NOTES

- *A 30-year-old lady was diagnosed to have acute appendicitis with classical features—pain, fever, vomiting and tenderness in the McBurney's point. A gynaecological examination revealed pelvic infection. An infected copper T was removed which was the cause of abdominal pain.*
- *A 22-year-old man underwent appendicectomy for right-sided abdominal pain. At laparotomy, appendix was normal. However, it was removed. He continued to have abdominal pain. An ultrasound of the abdomen revealed torsion of the undescended testis.* ***Nobody had examined his external genitalia!***
- *A 36-year-old male who had previous history of abdominal pain underwent appendicectomy for tenderness and rebound tenderness in the right iliac fossa. Operative surgery notes said that the appendix was slightly inflamed and seropurulent fluid was present in the right iliac fossa. After 2–3 days, greenish fluid (bile) started draining out through the tube. The condition of the patient deteriorated and on re-exploration this time, by midline incision revealed perforated chronic duodenal ulcer!!*

Complications of acute appendicitis

1. **Rupture** of appendix causes generalised peritonitis with 10–20% mortality rate. The treatment involves emergency laparotomy, appendicectomy and peritoneal wash followed by drainage of peritoneal cavity.
2. **Appendicular mass** (Figs 33.16 and 33.17)
   - Following an attack of acute appendicitis, infection is sealed off by greater omentum, caecum, terminal ileum,

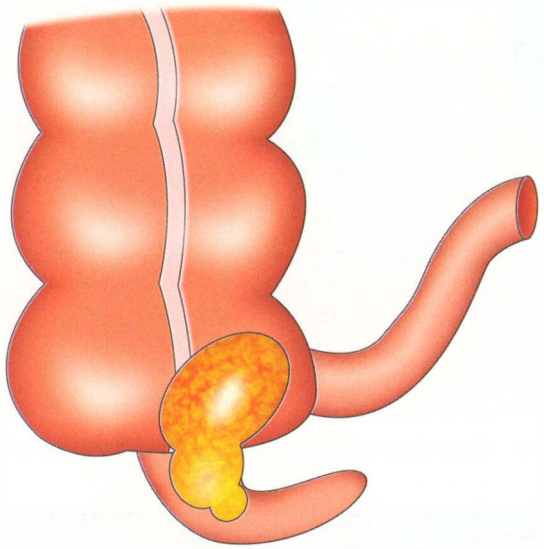


Fig. 33.16: Appendicular mass—tender, diffuse mass

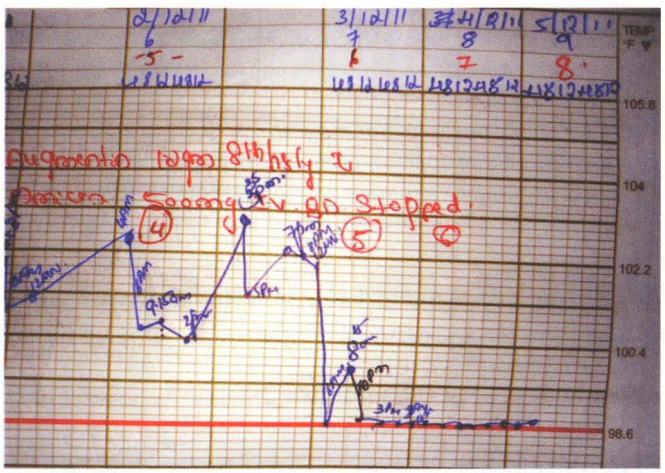


Fig. 33.17: Appendicular abscess—once drained, fever touched the base

CLINICAL NOTES

- *A 60-year-old lady was diagnosed to have appendicular mass and was undergoing conservative management. On the fourth day, she developed features of early septic shock. As the patient was not improving, laparotomy was done. It was a case of* ***volvulus of the caecum****.*

etc. which results in a tender, soft to firm mass in the right iliac fossa.

- **Presence of a mass** is a **contraindication for appendicectomy** because it is very difficult to remove appendix from such a mass. An attempt to remove it may result in a faecal fistula.
- It is treated by **Ochsner and Sherren regime**.
  - **A**spiration with Ryle's tube to give rest to the gut.
  - **B**owel care—purgatives should not be used (may cause perforation).
  - **C**harts—temperature, pulse, respiration, diameter of the mass. Swinging temperature, and increase in size of mass indicates an appendicular abscess.
  - **D**rugs to cover all the organisms—gram-positive, gram-negative and anaerobic organisms.
  - **E**xploratory laparotomy should not be done. However, when the condition of the **patient is not improving,** there is a suspicion of an abscess (Fig. 33.17) and when doubtful of the diagnosis, exploration is indicated (see the clinical notes above).
  - **F**luids (*see* Table 33.2)

  Patient is kept nil orally for a few days. During this time, intravenous fluids are given to correct dehydration.
- After 3–4 days, the abdomen becomes soft, tenderness decreases and once stools are passed, Ryle's tube is removed. Clear oral fluids followed by soft diet is given.

Table 33.2 Wisdom/mistakes/surprises for surgeon while conducting appendicectomy

Observes straw-coloured fluid	Observes bile-coloured fluid	Observes 'foul' fluid	Observes haemorrhagic fluid
Completes appendicectomy	Completes appendicectomy	Completes appendicectomy	Completes appendicectomy
Closes the wound	Puts a drain	Postoperative faecal fistula	Ignores fluid
Realises 3 days later, it was acute pancreatitis	Closes the wound	Re-explores by midline incision	Patient continues to have pain
How could it have been avoided?	Postoperative biliary fistula	Perforated Meckel's diverticulum	OBG consultation given
Serum amylase, lipase were not sent. A preoperative ultrasound was not done.	Asks for contrast CT, **realises duodenal ulcer perforation**, explores, sutures the perforation	Resection, anastomosis	Twisted ovarian cyst
Surgical wisdom: If these investigations were done before surgery, they could have helped the surgeon. Luckily, the patient recovered from this unnecessary, avoidable surgery.	He had not done a simple chest X-ray or plain X-ray abdomen erect preoperatively in this case	**Surgical wisdom:** Surgeon had not examined the terminal 2 feet ileum during appendicectomy	Laparotomy and ovariectomy **Surgical wisdom:** Surgeon had not done ultrasound and gynaecological consultation was not requested before surgery.

By one week, the patient is back to normal. **After 6–8 weeks**, patient is advised **elective appendicectomy**.

3. Perforated appendicitis

- Incidence is about 8–10%.
- More common in children and elderly patients.
- Delay in seeking medical treatment is the main factor.
- Other factors which precipitate perforation are diabetes mellitus, AIDS, faecolith.
- The pain usually localises to the right lower quadrant if the perforation has been walled off by surrounding intra-abdominal structures including the omentum.
- Diffuse pain in cases of generalised peritonitis.
- Rigors and chills with fever of 102°F (38.9°C) or above.
- As a complication of perforation peritonitis, portal pyaemia (pylephlebitis) can develop, it can be very dangerous.
- Emergency laparotomy, appendicectomy, drainage of pus, peritoneal lavage, antibiotics
- Mortality in these cases can be high.

4. Appendicular abscess (Fig. 33.18): If the infection is not controlled properly following an attack of appendicitis, an abscess can occur in relation to the appendix. They are (A) retrocaecal, (B) postileal and preileal, (C) pelvic, (D) subcaecal abscesses. Clinically, it presents with high-grade fever with chills and rigors and a tender boggy swelling in the right iliac fossa or in the right lumbar region. Pelvic abscess presents with diarrhoea. Diagnosis is by late presentation to the hospital (3–4 days) and high-grade fever with chills and rigors (Key Box 33.7).

A. Retrocaecal abscess is drained by extraperitoneal approach. An incision of 5 to 6 cm is made in the right iliac fossa and all muscles are divided. However, perito-neum is not opened. It is swept medially and pus is drained outside. Appendicectomy is done at a later date (Fig. 33.19).

B. Preileal and postileal abscesses are drained by a laparotomy.

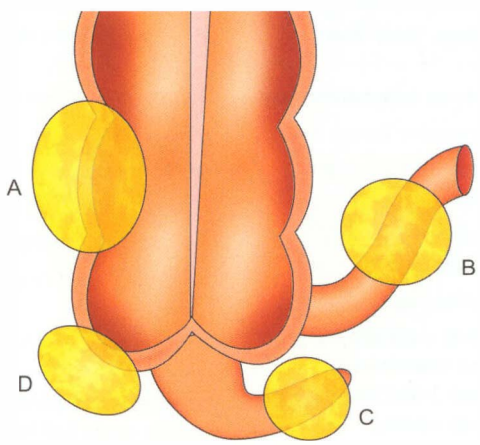


Fig. 33.18: Appendicular abscess (*see* text for A to D)

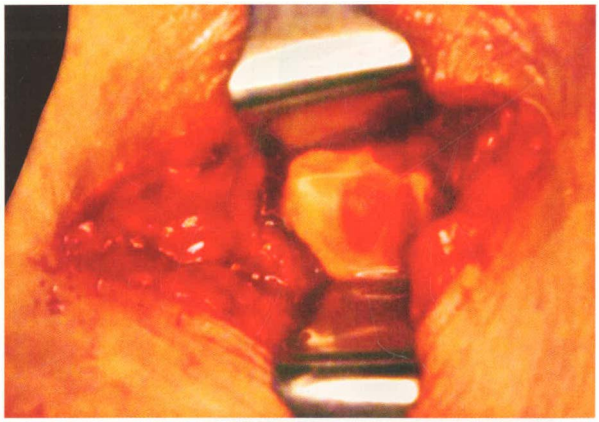


Fig. 33.19: Appendicular abscess is drained by extraperitoneal route

KEY BOX 33.7

APPENDICULAR ABSCESS

- Ultrasound/CT scan is done to assess the size and location of abscess
- Abscess greater than 4–6 cm in size needs to be drained by guided percutaneous aspiration or drainage through rectum or vagina
- Ongoing inflammation may force a surgeon to do appendicectomy open/laparoscopic at the same admission
- Those who improve require appendicectomy after 6 weeks

C. Pelvic abscess is drained *via* the rectum (*see* page 647)

D. Lumbar abscess (perinephric abscess) is drained through a loin incision.

Preoperative resuscitation

- Once diagnosis of acute appendicitis is suspected, the patient is admitted to the hospital.
- IV fluids—isotonic saline or Ringer lactate is given.
- Electrolytes are corrected especially in late cases of acute appendicitis/perforation peritonitis, etc.
- Ryle's tube is not necessary in simple appendicitis but is definitely required in complicated cases (peritonitis).
- Second generation cephalosporins along with metro-nidazole is given.
- Informed consent is taken.

Treatment

- **Emergency appendicectomy:** Emergency appendicectomy is offered when patient comes within 24 to 48 hours of abdominal pain. It is very important to rule out or detect a mass, especially if a decision is made to operate around 2nd or 3rd day. If a mass is palpable, it is better not to operate at that time (please refer to operative surgery, appendicectomy). A few important steps are given here (Figs 33.20 to 33.24).
- The appendix is identified by tracing *Taenia coli* which converges onto the base of the appendix. Mesoappendix is

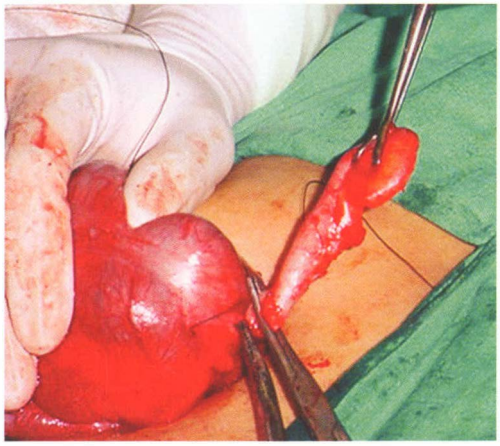


Fig. 33.20: Emergency appendicectomy—base is crushed

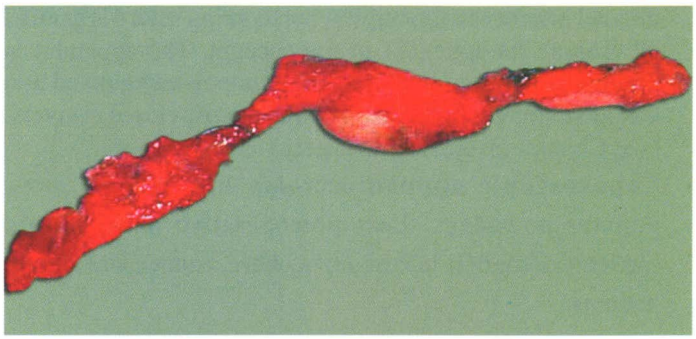


Fig. 33.21: Large faecolith resulting in acute appendicitis

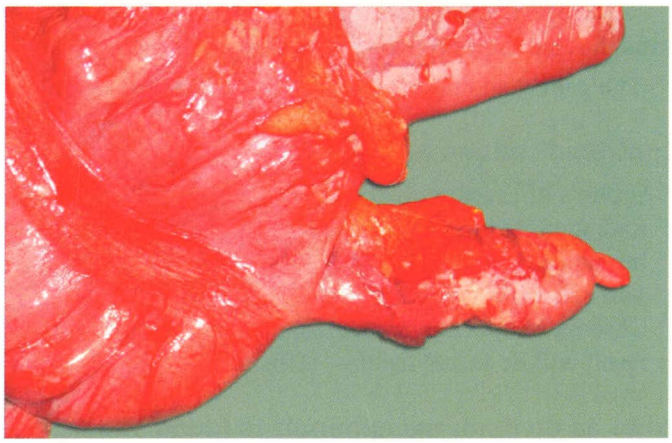


Fig. 33.22: Inflamed appendix at surgery (*Courtesy:* Dr Prasad, S. Professor of Surgery, KMC, Manipal)

Fig. 33.23: Appendicular perforation with abscess—appendicectomy could be done (*Courtesy:* Dr Annappa Kudva, Professor of Surgery, KMC, Manipal)

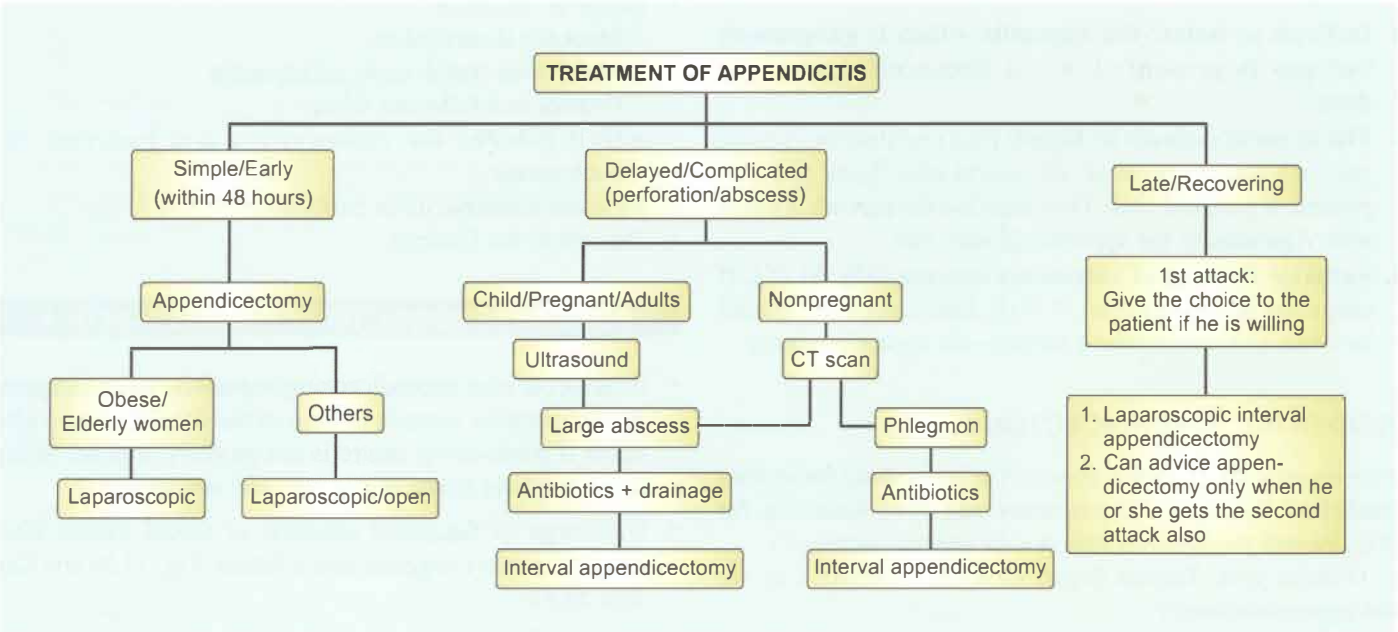


Fig. 33.24: Algorithm of treatment of appendicitis

divided in between ligatures. A purse-string suture is applied all around the appendix in the caecum. The appendix is divided in between ligatures, the stump is invaginated and the purse string is tightened. The abdomen is closed in layers. (*see* Chapter on operative surgery)

- **Laparoscopic appendicectomy** has become more popular nowadays. Less postoperative pain, speedy recovery. Benefit is maximum in obese, women and elderly patients.

PEARLS OF WISDOM

When you trace Taenia coli and you are not getting appendix, it means you are holding and tracing Taenia of the sigmoid colon. (It can be found on the right side sometimes.)

Problems encountered during appendicectomy

1. **The incision is small:** Location is higher up—never hesitate to close the incision and a midline incision is given and do appendicectomy. An attempt to remove the appendix with traction and limited exposure through McBurney incision may result in faecal fistula.
2. **Normal appendix is found:** Remove the appendix. Otherwise it may cause confusion to the next surgeon when this patient presents with abdominal pain. However, look for Meckel's diverticulitis, intestinal obstruction, stricture, etc.
3. **Gangrenous appendix involving base:** Problem one can face here is that the purse string can be applied but invagination of the stump is not possible. Risk of faecal fistula is also present. Appendicectomy, wash and a drain is kept.
4. **Difficult to isolate the appendix which is gangrenous but pus is present:** Limited ileocecectomy can be done.
5. **The appendix cannot be found:** First mobilise the caecum and look for subcaecal or retrocaecal sites. Look also into preileal or postileal sites. Then mobilise the ascending colon also. Agenesis of the appendix is very rare.
6. **Surprise findings of carcinoma caecum** (Fig. 33.25): If suspicion of a carcinoma is high, hemicolectomy should be done. Otherwise take a biopsy—do appendicectomy.

INCIDENTAL APPENDICECTOMY

It means removal of normal appendix at laparotomy for another condition. Examples: Laparotomy and ileal resection for stricture and anastomosis (can we do appendicectomy?).

Ovarian cyst: Torsion (right) ovary is removed. Can we add appendicectomy?

Since benefits of appendicitis/appendicectomy is more in young patients, if patient is under 30 years, it may be justifiable

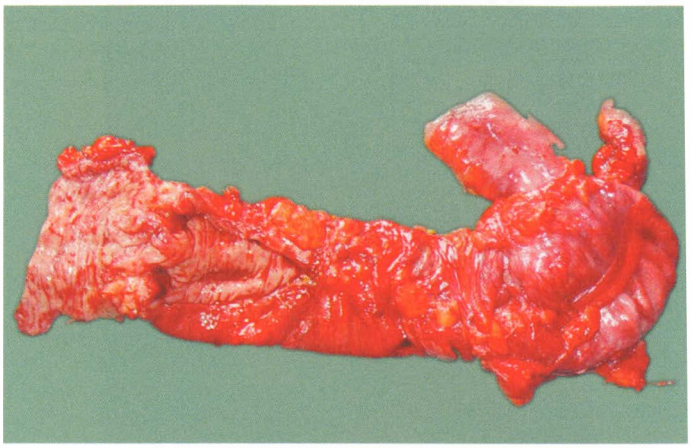


Fig. 33.25: Carcinoma colon with appendicitis

to do incidental appendicectomy provided it can be removed through same incision, without much difficulty.

The patient should be stable to tolerate the procedure.

Contraindications for incidental appendicectomy

- Crohn's of caecum
- Radiation treatment of caecum
- Immunosuppression
- Vascular grafted patient (aortoiliac, etc.)
- Chances of infection are high in this group of patients. The result will be faecal fistula—difficult to treat.

What to do if normal appendix is found at surgery?

- Normal white appendix is called Lily-white appendix.
- It is removed because the 'scar' should not add confusion later to a doctor whether appendix was removed or not.
- However, examine:
  - Meckel's diverticulum
  - Mesenteric lymph node enlargement
  - Ovaries and fallopian tubes
  - Gall bladder for cholecystitis and pancreas for pancreatitis.
- Rule out duodenal ulcer perforation.
- Document the findings

POST-APPENDICECTOMY FAECAL FISTULA

- It can occur after appendicectomy especially when gangrene of the appendix extends to base of the caecum. It can also occur if purse-string suture is not properly applied, injury to the terminal ileum or caecum, etc. occurs.
- Discharge of faeculent contents or faecal matter after appendicectomy suggests faecal fistula (Fig. 33.26 and Key Box 33.8).
- Usually discharge stops after a few days provided there is no distal obstruction.

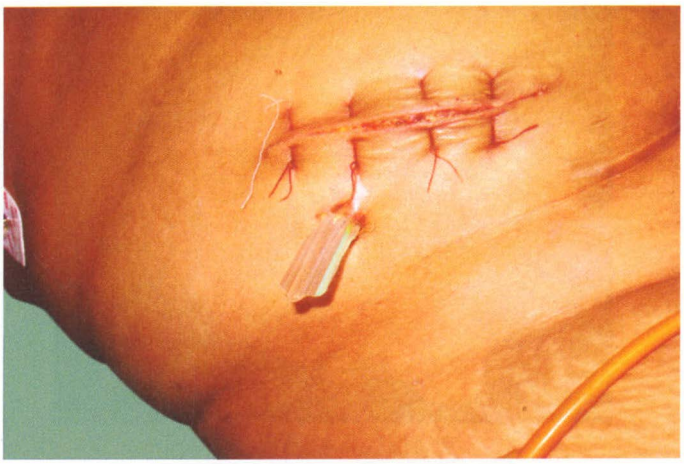


Fig. 33.26: This patient had faecal fistula which healed after two weeks of conservative management

KEY BOX 33.6

FAECAL FISTULA—CAN OCCUR

- After drainage of appendicular abscess
- After appendicectomy—if purse string sutures are not properly applied
- If the caecum is also involved by inflammation
- If the cause of appendicitis is carcinoma
- If chronic diseases develop or are present—tuberculosis, Crohn's or actinomycosis
- If appendicitis is associated with carcinoma caecum

- Cases which do not respond to conservative treatment are managed by resection of the diseased portion of the caecum or ascending colon.

PEARLS OF WISDOM

The important causes for faecal fistula are carcinoma caecum and ileocaecal tuberculosis in India and Crohn's disease in the West.

NEOPLASM OF THE APPENDIX

1. Carcinoid tumour

- It is the most common neoplasm of the appendix, less aggressive, majority are benign and cured with simple appendicectomy (*see* Chapter 28 for more details).
- Goblet cell carcinoid tumour—it is more aggressive, requires right hemicolectomy if the tumour is more than 2 cm, has more than 2 mitosis per high power field and lymphovascular invasion, adenocarcinoma of the appendix.

2. Carcinoma

- It is very rare.
- Often it is colonic type. Other type is mucinous adenocarcinoma.
- Can present as acute appendicitis due to obstruction caused by the tumour.
- Mucinous variety has better prognosis.
- Colonic variety should be treated by right hemicolectomy.

3. Cystic neoplasm of the appendix

- Rare occurrence
- Simple cyst (non-neoplastic mucocoele) and mucinous cystadenoma (like pancreatic).
- Can attain large size.
- Diagnosis is by ultrasound/CT scan.
- Appendicectomy is the treatment of choice.
- It can rupture into peritoneal cavity resulting in pseudomyxoma peritonei.

MUCOCOELE OF THE APPENDIX

Definition: It means accumulation of mucus within the lumen of the appendix.

Causes: It can be a simple retention cyst due to blockage by foreign body or mucosal hyperplasia. It can also be due to a mucinous adenocarcinoma (Fig. 33.27).

Pathology (Figs 33.28 to 33.31)

- The majority of epithelial tumours of the appendix are mucin rich, thus results in gross distension.
- Mucocoeles resulting from non-neoplastic occlusion (simple retention cysts) rarely exceed 2 cm in diameter.
- Mucinous neoplasms of the appendix are by far the most common cause of mucocoeles.
- Mucocoeles larger than 2 cm are more likely to represent benign neoplasms.

Diagnosis

- It is impossible to differentiate clinically mucocoele of the appendix and acute appendicitis when they present with abdominal pain. If a mass is palpable, it can be confused with appendicular mass.

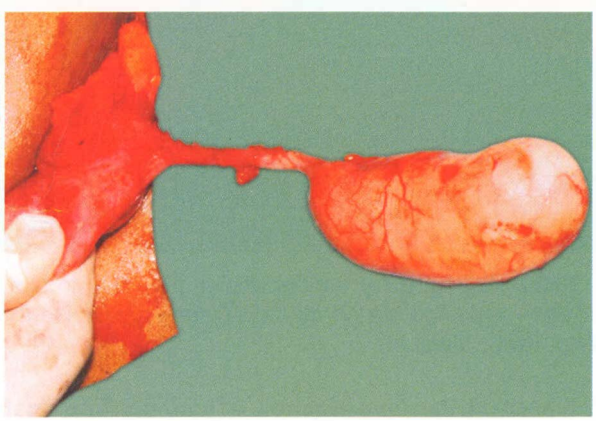


Fig. 33.27: Mucocoele of the appendix (*Courtesy:* Dr Raghunath Prabhu, KMC, Manipal)

Fig. 33.28: Large mucocoele removed along with caecum and terminal ileum

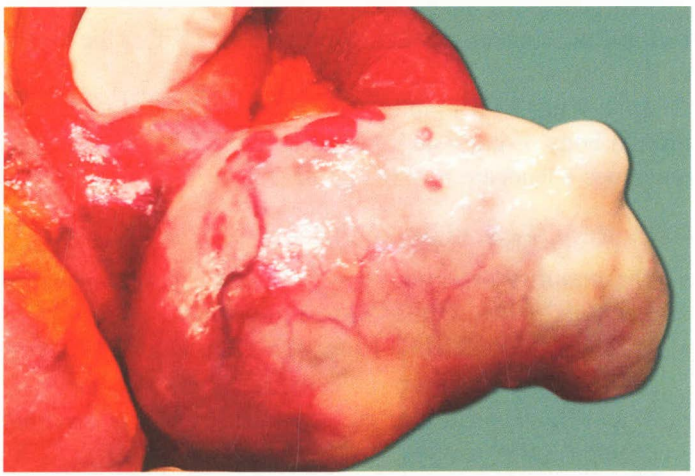


Fig. 33.29: Mucocoele (*Courtesy:* Dr Rajesh Sisodia, KMC, Manipal)

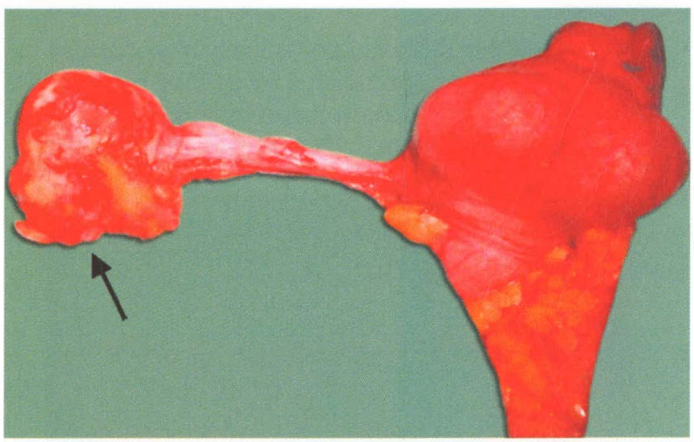


Fig. 33.30: Appendicular tumour arising from the tip has been removed—reported as carcinoid (*Courtesy:* Dr Raghunath Prabhu, KMC, Manipal)

- CT scan is the investigation of choice. The anatomic relationship between the elongated cystic mass and the caecum is usually more clear at CT scan than at ultrasound.

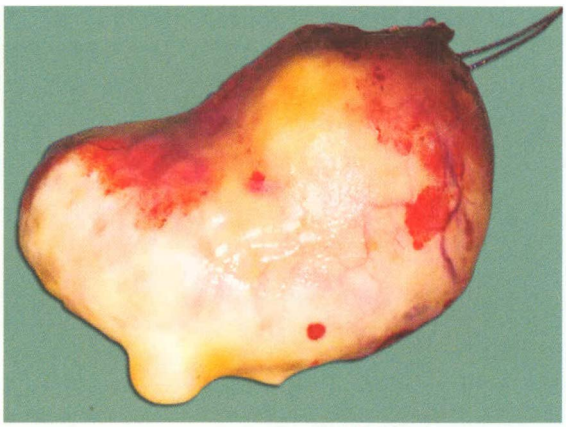


Fig. 33.31: Mucocoele specimen

Complications

1. **Gross enlargement** and can present as mass abdomen.
2. **Rupture** will result in pseudomyxoma peritonei (more details are given in Chapter 27, Peritonitis).
3. **Secondary infection:** Can result in 'empyema' of the appendix.

PEARLS OF WISDOM

Rule out adenocarcinoma of the base of the appendix causing mucocoele.

Treatment: Appendicectomy

MISCELLANEOUS

VALENTINO APPENDIX

- Rudolf Valentino was an Italian actor acting in Hollywood who was operated for right iliac fossa pain with features of peritonitis in the early 20th century. Following a few days of surgery, he died of sepsis. The actual disease was perforated duodenal ulcer. This is a typical case scenario that holds true even today. The contents gravitate down along right paracolic gutter. The symptoms and signs mimic appendicitis. CT scan is the investigation needed to rule out other causes (*see* page 637).

POST-APPENDICECTOMY SEPSIS (A case report)

A 32-year-old man presented to casualty with septic shock after 5 days after appendicectomy. It was a difficult appendicectomy. Gangrene of the appendix was almost involving the base.

On examination he was having paralytic ileus and jaundice. Abdomen was distended—guarding was present, more in the right iliac fossa. He was admitted to the hospital.

Investigations

- Total counts were 20,000 cells/cu mm
- Urea: 51, Cr: 1.1

- K: 3.2, Na: 129
- TB: 19 mg/DI, DB: 16.6, ALP: 167
- AST: 189 units, ALT: 100 units

Remarks: It showed he was in sepsis—counts were elevated, urea was high—renal failure sets in slowly, increased bilirubin levels—sepsis with cholestasis.

Plain X-ray chest revealed free gas under the diaphragm (Fig. 33.32).

CT scan: Done after hydration—showed pneumoperitoneum and liver cyst (incidental) and free fluid in the peritoneal cavity (Fig. 33.33).

Conclusion

He was in sepsis. The reason was probably that the appendicular base (stump) had given way.

Exploratory laparotomy

- Faecal peritonitis
- One litre of frank purulent pus in the peritoneal cavity.
- Gangrene of lateral wall of caecum with sloughing of caecal wall.
- Appendix not seen—post-appendicectomy
- Ileum normal
- Cystic lesion on anterior surface of right lobe of liver
- Rest of viscera are normal

Procedure

- Limited resection of ileocecal segment and end-to-end ileo-ascending single layer anastamosis
- Peritoneal lavage
- Drains in the pelvis and subhepatic space
- Skin not closed (wound infection is very common)

Postoperative

6th postoperative day

- Patient had greenish discharge from the right DT
- Anastomotic leak and enterocutaneous fistula was suspected
- Patient was passing flatus
- RS: Basal crepitations
- Managed conservatively
- TPN was given for 5 days
- Discharge subsided by 5 days

8th postoperative day

- Breathlessness
- Fever
- Hypoxia: SpO_2: 85%
- Chest X-ray—pneumonia (Fig. 33.34)
- Intubated, ventilated for 5 days, appropriate antibiotics
- By 20th day, he was discharged from the hospital—leak had stopped.

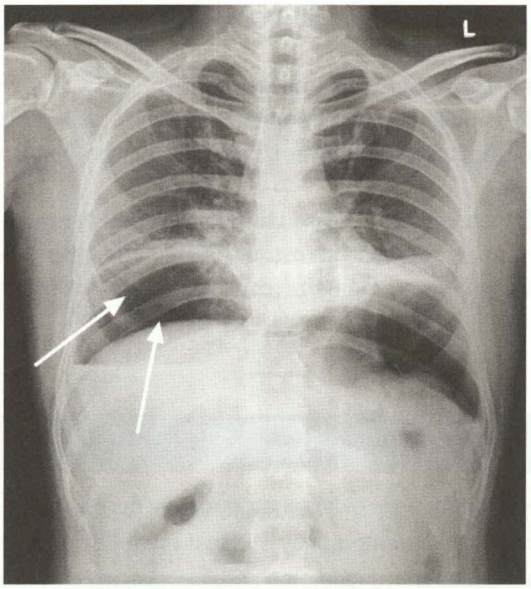


Fig. 33.32: Free gas under the right dome of the diaphragm

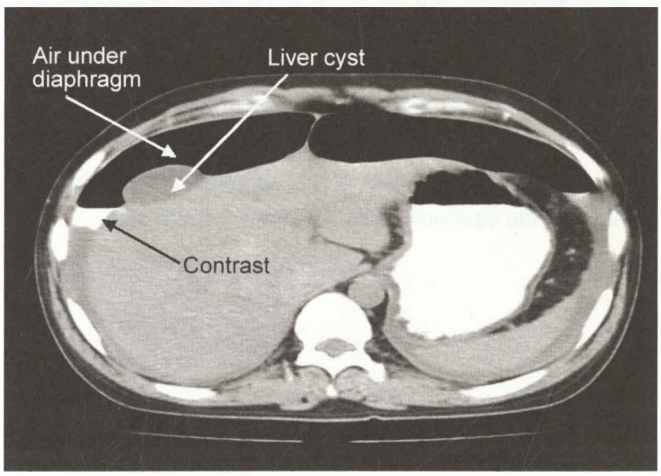


Fig. 33.33: Free gas under the right dome of the diaphragm pneumoperitoneum and liver cyst

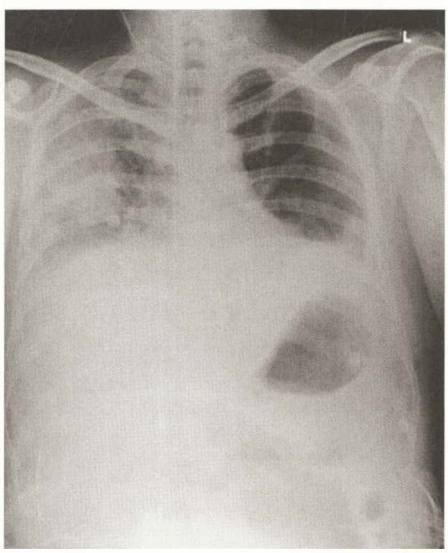


Fig. 33.34: Right lung pneumonia

- This case report has been given here for the following message

1. Acute appendicitis can be dangerous
2. Leak should be suspected if a patient who underwent appendicectomy does not improve in the postoperative period.
3. High total count, increased bilirubin, oliguria suggest sepsis
4. CT scan is the best investigation in such cases. When in doubt reexplore. Danger lies in delay, not in resurgery.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated with flowcharts and coloured pictures.
- New algorithm of treatment of appendicitis and a Wisdom Table 33.2 have been added.
- Valentino appendix and neoplasms of the appendix have been added.
- Multiple choice questions have been added.

INTERESTING 'MOST COMMON'

- Most common surgical emergency encountered by a general surgeon is acute appendicitis.
- Most common emergency surgical operation is appendicectomy
- Most common nonobstetric surgical disease of the abdomen during pregnancy is acute appendicitis.
- Most significant symptom of acute appendicitis is migratory pain.
- Most significant sign of acute appendicitis is rebound tenderness in the McBurney's point.
- Most prominent scoring system to diagnose acute appendicitis is Alvarado score.
- Most common anaerobic bacteria in acute appendicitis is *Bacteroides fragilis* and aerobic bacteria is *Escherichia coli.*
- Most common complication after appendicectomy is wound infection.
- Most common age group for acute appendicitis is below 40 years.
- Most common neoplasm of the appendix is carcinoid tumour.
- Most of the carcinoids are less than 1 cm in size.

MULTIPLE CHOICE QUESTIONS

1. The most common position of the appendix is:

A. Subhepatic B. Subcaecal
C. Retrocaecal D. Pelvic

2. The incidence of appendicitis is less after 30 years because:

A. The appendix undergoes involution
B. The lymphatic tissue in the appendix decreases
C. Most people would have had their appendices removed
D. The vascularity reduces

3. The name Sheshachalam is associated with which of the following arteries?

A. Accessory appendicular artery
B. Appendicular artery
C. Ileocolic artery
D. Posterior caecal artery

4. Appendicular orifice is occasionally guarded by an indistinct semilunar fold of mucous membrane called:

A. Valve of Gerlach B. Valve of Heister
C. Valve of Kerckring D. Valve of Houston

5. The most common scoring system used for appendicitis is _____ scoring system.

A. Child-Pugh B. Furtado
C. Murray D. Alvarado

6. Palpation of left iliac region of abdomen produces pain in the right iliac region in appendicitis because of:

A. Sympathetic reaction
B. Displacement of colonic gas and small bowel coils
C. Sigmoid colon is also affected
D. Ileocolic reflex

7. Cope's psoas test is positive in:

A. Retrocaecal appendicitis
B. Pelvic appendicitis
C. Preileal appendicitis
D. Subcaecal appendicitis

8. Rebound tenderness in acute appendicitis is called:

A. McBurney's sign B. Blumberg's sign
C. Rovsing's sign D. Sherren's sign

9. The most common cause of nonobstetric emergency with abdominal pain in pregnancy is due to:

A. Acute appendicitis B. Acute cholecystitis
C. Acute gastritis D. Acute hepatitis

10. Contraindications for incidental appendicectomy include all of the following *except:*

A. Crohn's of caecum
B. Radiation treatment of the rectum
C. Immunocompetent individuals
D. Previous vascular reconstruction in the abdomen

11. The following statement is TRUE about appendicular abscess:

A. Abscess greater than 4–6 cm in size needs to be drained by laparotomy
B. Appendicectomy must be done along with laparotomy for appendicular abscess
C. Can present with diarrhoea
D. Conservative management is advised till inflammation settles down.

12. Most common aerobic bacteria involved in acute appendicitis is:

A. *Samonella typhi* B. *Streptococcus*
C. *Escherichia coli* B. *Clostridium perfringens*

13. The following statement is FALSE about occurrence of faecal fistula following appendicectomy:

A. Faecal fistula can occur if the cause of appendicitis is carcinoma caecum
B. Faecal fistula can occur if chronic diseases such as tuberculosis is present
C. Faecal fistula can occur if purse string sutures are not applied properly
D. It is always due to actinomycosis

14. The most reliable symptom of acute appendicitis is:

A. Fever B. Migratory pain
C. Right iliac fossa pain D. Vomiting

15. Appendicular perforation is common because of the following reasons *except:*

A. Appendix is a cul-de-sac
B. It has blood supply with profuse collaterals
C. It has a narrow lumen
D. The muscle coat of appendix is thin

ANSWERS

1 C	2 B	3 A	4 A	5 D	6 B	7 A	8 B	9 A	10 C
11 C	12 C	13 D	14 B	15 B					

34

Hernia

- Anatomy
- Indirect hernia
- Direct hernia
- Clinical examination
- Complications
- Recurrent hernia
- Special hernias
- Giant hernia
- Sportsmen hernia
- Sliding hernia
- Femoral hernia
- Rare types of femoral hernia
- Umbilical hernia
- Incisional hernia
- Epigastric hernia
- Interparietal hernia
- Spigelian hernia
- Lumbar hernia
- Obturator hernia
- Perineal hernia
- What is new?/Recent advances

Introduction

- Hernia is a common condition affecting patients, especially inguinal hernia in males and incisional hernia in females. Even though several types of surgery have been described for hernias, mesh hernioplasty remains the gold standard treatment. Majority of the hernias require surgical treatment leaving apart small asymptomatic direct hernias in elderly. Today, laparoscopic hernia is becoming gold standard. Obstructed hernia is an emergency and late cases carry significant mortality. As far as students are concerned, hernia is the most common case in the examination. Hence, detailed clinical examination of hernia, complications, and various types of hernias and their treatment have been described in this chapter.
- Hernia means to bud or protrude or rupture (Latin).

Definition

Abnormal protrusion of a viscus or a part of it through a weak point in the body (opening) is known as a hernia. Inguinal hernia occurs either through the deep inguinal ring (indirect hernia) or through the posterior wall of the inguinal canal (direct hernia).

ANATOMY OF THE INGUINAL REGION (Fig. 34.1)

Inguinal ligament (Poupart's ligament) (1)

- It is the ligamentous portion of the external oblique aponeurosis which folds inwards and extends from anterior superior iliac spine to the pubic tubercle.
- The midpoint between these two structures is called midpoint of the inguinal ligament.

Lacunar ligament (Gimbernat's ligament)

- Some fibres of inguinal ligament pass posteriorly to attach to superior pubic ramus lateral to the tubercle and forms lacunar ligament.
- The midpoint between the anterior superior iliac spine and pubic symphysis is called midinguinal point.

Inguinal canal (2)

- It is 4 cm in length extending from the deep inguinal ring to the superficial inguinal ring.

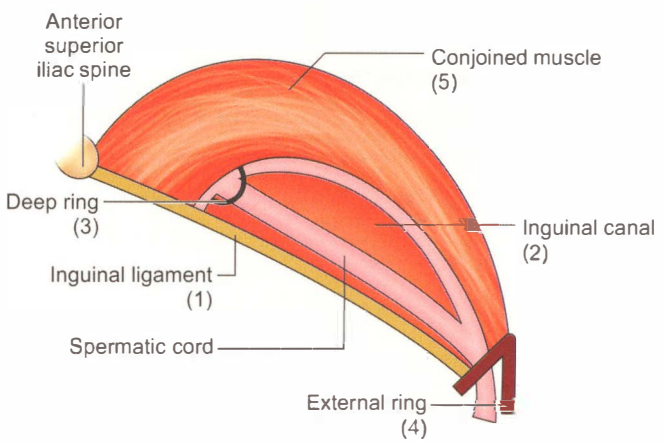


Fig. 34.1: Anatomy of the inguinal region

Deep ring (internal ring) (3)

It is a 'U' shaped defect in the fascia transversalis which forms the posterior wall of the inguinal canal. It lies 1.25 cm above the midpoint of the inguinal ligament.

External ring (superficial ring) (4)

Superficial ring is a triangular defect in external oblique aponeurosis. It is bounded by the lateral and medial crura formed by the external oblique aponeurosis and the base of the triangle is formed by the pubic crest.

Boundaries of inguinal canal

- **Anterior:** External oblique aponeurosis and a few fibres of the **conjoined muscle** (especially of internal oblique) laterally.
- **Superior:** Arched fibres of the **conjoined muscle** (5).
- **Inferior:** Inguinal ligament and the lacunar ligament on the medial side (Gimbernat's ligament).
- **Posterior:** Fascia transversalis and the conjoined tendon medially. Thus, the inguinal canal is strong in the lateral part anteriorly and the medial part posteriorly.

Contents of inguinal canal

1. Spermatic cord (Key Box 34.1)
2. Ilioinguinal nerve (Key Box 34.2)
3. Genital branch of genitofemoral nerve
4. Round ligament in females
5. Vestigeal remnant of processus vaginalis sac.

Myopectineal orifice of Fruchaud (Fig. 34.2)

- This weak area is the site of all groin hernias according to Fruchaud.
- It is the area between inguinal ligament anteriorly and iliopubic tract posteriorly.
- **Iliopubic tract:** It is the thickened inferior margin of the transversalis fascia which appears as a fibrous band running parallel and posterior (deep) to inguinal ligament. It inserts into superior pubic ramus to form lacunar ligament.

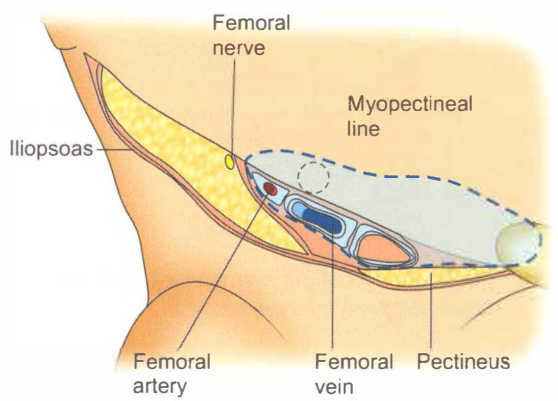


Fig. 34.2: Myopectineal line and anatomy

KEY BOX 34.1

CONTENTS OF THE SPERMATIC CORD

- Vas deferens
- Testicular artery
- Artery to the vas
- Cremasteric artery
- Pampiniform plexus of veins
- Lymphatics
- Sympathetic nerves
- Genital branch of genitofemoral nerve
- Processus vaginalis

KEY BOX 34.2

ILIOINGUINAL NERVE

- The ilioinguinal nerve is a branch of the first lumbar nerve (L1). It separates from the first lumbar nerve along with the larger iliohypogastric nerve.
- Ilioinguinal nerve does not pass through the deep inguinal ring. It only travels through part of the inguinal canal.
- After going through inguinal canal, it pierces the internal oblique muscle, distributes nerve fibres to it, and then accompanies the spermatic cord through the superficial inguinal ring.
- Divides into anterior scrotal nerve and anterior labial nerve.
- Supplies the skin of the upper and medial part of the thigh, scrotum and vulva.
- Entrapment or injury to the ilioinguinal nerve is one of the causes of postherniorrhaphy pain.
- Hence, a few recommend division of ilioinguinal nerve during hernia surgery.

- **Boundaries of myopectineal orifice of Fruchaud:**

  Superiorly: Arched fibres of internal oblique

  Laterally: Iliopsoas muscle

  Medially: Lateral border of rectus abdominus muscle

  Inferiorly: Pubic pecten—Cooper's ligament

- **Surgical importance of iliopubic tract:** Recognition of this is a part of laparoscopic repair (initial step)—visualising from within. This structure reinforces posterior wall and floor of inguinal canal as it bridges structures traversing subinguinal space.

Hesselbach's triangle (Fig. 34.3)

- It is bounded medially by lateral border of rectus abdominus muscle, laterally by inferior epigastric artery and inferiorly by inguinal ligament.
- Direct hernias occur commonly through Hesselbach's triangle (medial), indirect hernia lateral to inferior epigastric artery.

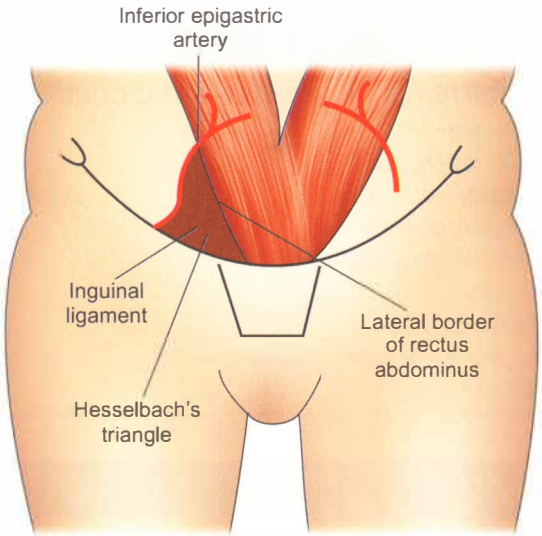


Fig. 34.3: Hesselbach's triangle

INGUINAL DEFENCE MECHANISMS

1. Obliquity of inguinal canal (in children it is straight).
2. During straining or coughing, the conjoined tendon contracts, and since it forms the anterior, superior and posterior boundaries, it closes the inguinal canal—**shutter or sphincter-like effect**.
3. Increased intra-abdominal pressure produces plugging effect at the external ring. The deep ring is pulled upwards and laterally because it is adherent to the posterior surface of **transversalis muscle**. This occludes the ring and prevents herniation—**Ball valve** effect.

CLASSIFICATION OF HERNIA

I. The European hernia society classification:

- Primary (P), Recurrent (R)
- Lateral (L), Medial (M), Femoral (F)
- Defect size assumed to be 1.5 cm

Thus primary direct hernia with 3 cm defect size is written as PM2.

II. Anatomical classification

1. Indirect hernia
2. Direct hernia

III. Nyhus classification

This classification is based **primarily on the defect**, which helps in planning an appropriate repair.

Type I: Indirect hernia with normal deep ring

Type II: Indirect hernia with dilated deep ring

Type III: Based on posterior wall defect

a. Direct
b. Pantaloon
c. Femoral

Type IV: Recurrent hernia.

IV. Gilbert's classification (Table 34.1)

- It is based on the defect in the posterior wall (direct hernia) or defect in the internal ring (indirect).
- Depending upon the defect, the suggested repair is given below. However, basic principles are the same.
- The last two types—Type VI and Type VII are modifications by Robbin.

Table 34.1 Gilbert's classification and suggested repair

Types	Classification	Repair
I.	Snug internal ring Preperitoneal indirect sac Does not admit one finger	Herniorrhaphy or hernioplasty
II.	Moderately enlarged internal ring Bubonocoele Admits one finger	Herniorrhaphy or hernioplasty
III.	Large defect—2 or 3 finger-breadths internal ring. May be sliding hernia	Preperitoneal mesh by slitting transversalis fascia
IV.	Large direct hernia with full blow out defect Internal ring is normal	Mesh repair
V.	Direct hernia with punched out hole/defect in the transversalis fascia The internal ring is intact	Plug the defect or purse-string closure of the defect followed by mesh repair
VI.	Pantaloon hernia	Mesh repair
VII.	Femoral hernia	Femoral hernia repair

AETIOLOGY OF HERNIA: WHAT CAUSES HERNIA?

- Indirect hernia occurs largely due to persistent processus vaginalis sac. Manifestations of this can be seen in elderly patients in whom an indirect hernia can be triggered by some factors which increase intra-abdominal pressure.
- Direct hernia occurs mainly due to weakness of transversalis fascia in Hesselbach's area. Increase in abdominal pressure can occur due to chronic cough, constipation or difficulty in passing urine, development of ascites (portal hypertension, nephrotic syndrome), etc.
- Collagen disorder: In prune-belly syndrome, collagen fibre disorder causes development of not only hernias but also intestinal hernias, bilateral hernias, etc. (Key Box 34.3).
- Hernia occur due to inherited imbalance in the types of collagen.

KEY BOX 34.3

CAUSES OF HERNIA

1. **Congenital**

   Persistent processus vaginalis sac: chief cause of indirect hernia.

2. **Collagen fibre disorder**
   - Prune-belly disorder—congenital
   - Smoking: Acquired collagen deficiency
3. **Obesity**
4. **Chronic causes of increased intra-abdominal pressure**
   - Chronic cough
   - Chronic constipation
   - Straining at micturition
   - Ascites
5. **Weakness of conjoined tendon/rupture of a few fibres**
   - Lifting heavy weight
   - Postappendicectomy—injury to ilioinguinal nerve.
   - Chronic illness/debilitating disease causing weakness of transversalis fascia in the Hesselbach's area.

INDIRECT HERNIA

It is a herniation of abdominal contents through the deep ring into the inguinal canal. **Indirect hernia occurs due to persistent processus vaginalis sac.** It is the most common type of hernia in the body. The preformed sac passes through the deep ring, traverses the inguinal canal and may extend into the scrotum through the external ring. As it comes into the inguinal canal, it is invested by the following coverings:

1. External spermatic fascia derived from external oblique aponeurosis.
2. Cremasteric fascia derived from internal oblique.
3. Internal spermatic fascia from fascia transversalis.

Processus vaginalis sac

- The testis originates in the lumbar region—in the retroperitoneum. Testis is guided or pulled down by a ligament called as gubernaculum. As the testis is pulled down it also pulls down the peritoneum along with it. This is processus vaginalis sac (tube). This process is obliterated in the normal situation. However, failure to close or obliterate is responsible for development of hernia.
- **New theory or recent advances:** Hormonal cause has been responsible for development of a hernia. Hepatocyte growth factor and calcium gene related peptide influence closure of the tube.

Parts of the hernia (Fig. 34.4)

Hernial sac is part of the peritoneum which is dragged into the inguinal canal. The mouth of the sac is in the peritoneal cavity. The neck is the narrowest portion (deep ring). The actual hernial sac has a body and a fundus. Depending upon the contents it can be named as follows: Omentum—omentocoele, Intestine– enterocoele, Littre's hernia—hernia containing Meckel's diverticulum. It may also contain ovary or appendix. **When part of the wall of the gut is involved, it is known as Richter's hernia.**

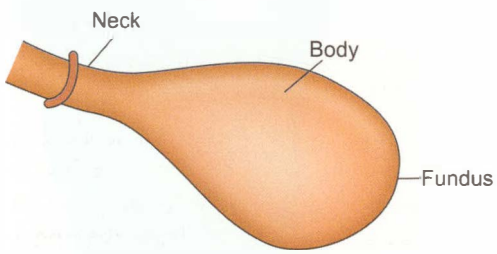


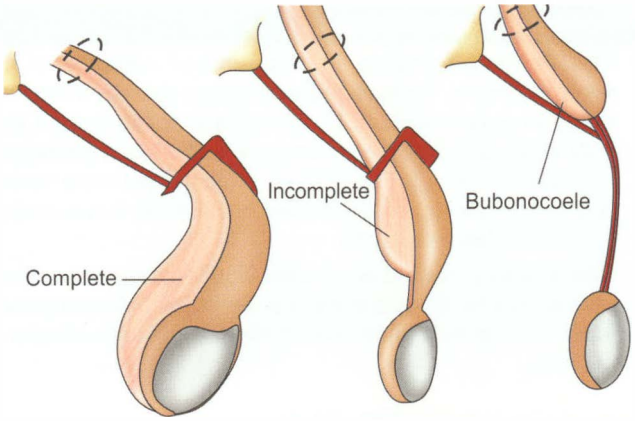
Fig. 34.4: Parts of the hernial sac

Types of indirect hernia

1. **Complete hernia** (scrotal): When the sac is patent up to the bottom of the scrotum, it is a **complete scrotal hernia** (Fig. 34.5).
2. **Funicular**: The processus vaginalis sac is patent up to the root of scrotum, it is a **incomplete indirect hernia** (Figs 34.6 and 34.8).
3. **Bubonocoele**: Processus vaginalis sac is confined to the inguinal region or the inguinal canal only. Such hernias are seen in young patients (Fig. 34.7).

Coverings of the indirect inguinal hernia from outside to inside

1. Skin
2. Two layers of superficial fascia: Fatty and membranous (Camper's and Scarpa's fascia respectively).
3. External spermatic fascia, a continuation of the external oblique aponeurosis.
4. Cremaster muscle and fascia, a continuation of the internal oblique.



Figs 34.5 to 34.7: Three types of indirect hernia

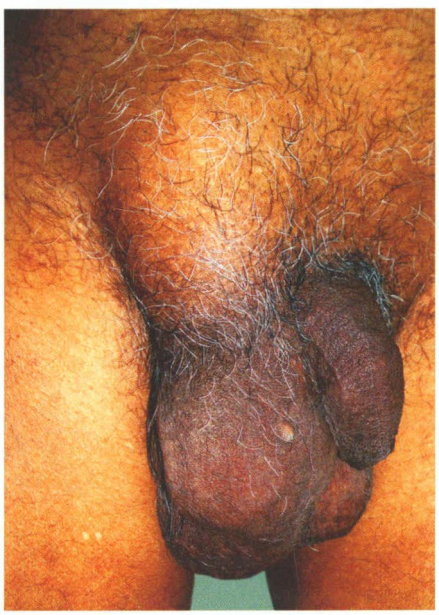


Fig. 34.8: Incomplete hernia bulge is seen

5. Internal spermatic fascia: Derived from the fascia transversalis.
6. Extraperitoneal fat
7. Peritoneum

DIRECT HERNIA

It is always acquired (Fig. 34.9). It occurs through ***Hesselbach's triangle,*** a weakness in the ***posterior wall*** of the inguinal canal (transversalis fascia). Its boundaries are:

- Medially: Lateral border of the rectus abdominis
- Laterally: Inferior epigastric artery
- Below: Inguinal ligament

Coverings of the direct inguinal hernia from outside to inside

1. Skin
2. Two layers of superficial fascia
3. External oblique aponeurosis

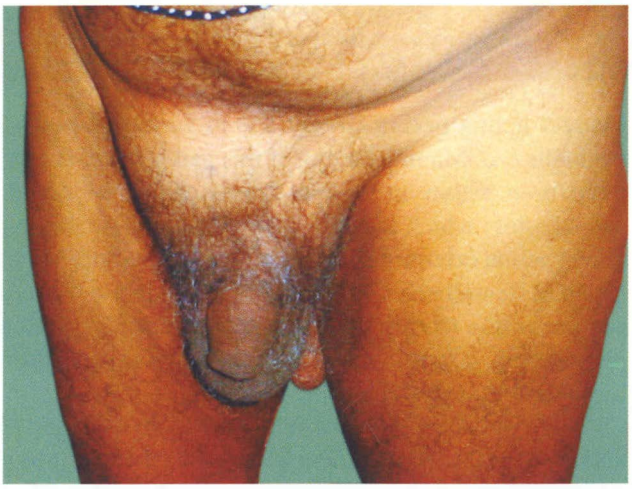


Fig. 34.9: Direct hernia: Pops out when patient stands

4. Conjoined tendon
5. Fascia transversalis
6. Peritoneum

Precipitating factors

1. Weakness of fibres of transversus abdominis or congenital absence of a few fibres is a major factor responsible for direct hernia.
2. In elderly patients, it is precipitated by:
   - Chronic cough, chronic bronchitis
   - Pops on coughing (Fig. 34.9)
   - Difficulty in passing urine due to benign prostatic hypertrophy (BPH).
   - Chronic constipation due to habitual constipation, or malignancy of left colon.
3. Smoking: Decreased strength of abdominal muscles due to decreased elastin.

PEARLS OF WISDOM

As the direct hernia pushes through the posterior wall, it is very unusual for it to descend into the scrotum.

Ogilvie hernia

This is a type of direct hernia wherein the hernial sac appears through a circular defect (congenital) in the conjoined tendon.

CLINICAL EXAMINATION OF A CASE OF HERNIA

History

- Swelling in the inguinal region which is gradually increasing in size.
- To start with, the swelling disappears on lying down and increases on straining, walking, etc. Later it cannot be reduced (due to adhesions).

- History of dragging pain indicates omentocoele.
- Since the omentum is attached to the stomach above and supplied by T10, the pain is referred to the umbilical region.
- Sudden, severe pain in the hernia, vomiting and irreducibility indicates 'obstructed hernia'.
- H/o chronic cough, constipation, difficulty in passing urine should be asked. If present, it may suggest the cause of hernia.

History of appendicectomy

Division of ilioinguinal nerve during appendicectomy may cause denervation of fibres of the right transversus abdominis, which forms 'U' shaped ring, resulting in weakness of the abdominal wall.

Inspection (a model case of incomplete hernia) (Fig. 34.10 and 34.11)

It should be done in the standing position. Both sides should be checked.

- There is a swelling in the inguinal region extending to the root of the scrotum measuring about 6 × 3 cm. **Its surface is smooth, borders are round and skin over the swelling is normal** and it is pyriform in shape.
- Ask the patient to cough—**expansile impulse on cough is present**. If peristalsis is present, it indicates an enterocoele. Expansile impulse on cough is diagnostic of hernia (Key Box 34.4).
- Presence of scar indicates a recurrent hernia. Ragged scar indicates infection.
- Direct hernia pops out as soon as the patient stands and often it is bilateral.

Palpation

- Inspectory findings should be confirmed.
- Swelling is soft, and gurgles if it is an enterocoele.
- It may be firm or granular if it is an omentocoele.

1. **Ask the patient to cough**—expansile impulse is felt at the root of scrotum.
2. **Getting above the swelling**[1] should be done in the standing position.

KEY BOX 34.4

EXPANSILE IMPULSE ON COUGH

- Hernia
- Meningocoele
- Dermoid cyst with intracranial communication
- Laryngocoele
- Lymphatic cyst in children
- Empyema necessitatis

Fig. 34.10: Incomplete indirect hernia

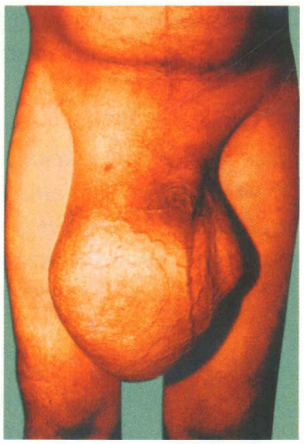


Fig. 34.11: Complete indirect hernia—scrotal hernia

- At the root of scrotum, the spermatic cord is palpated between the finger and the thumb. In cases of complete indirect hernia, spermatic cord cannot be felt as a naked structure because it is covered anterolaterally by the sac. This is called as **getting above the swelling not possible** (negative).

PEARLS OF WISDOM

Getting above the swelling is a test to differentiate scrotal swellings from inguinoscrotal swellings.

3. **Reducibility**—ask the patient to lie down.
- If the swelling becomes smaller or disappears, it is a hernia (hydrocoele is not reducible).
- **Omentocoele:** Initially, reduction is easy but later, becomes difficult (due to adhesions).
- If it is difficult to reduce, ask the patient to reduce it. Otherwise, flex and medially rotate the hip and try to reduce it, a method called as ***taxis***.
- If in spite of this, the swelling is not reduced, it is called as an irreducible hernia.

4. **External ring invagination test** (Fig. 34.12): At the root of the scrotum, skin is gathered and lifted up with the little finger. It is then invaginated into the external ring. As the external ring is stretched in indirect hernia, the finger goes obliquely and laterally. In a direct hernia, the finger goes backwards, and the superior ramus of the pubic bone can be felt as a bare bone. On asking the patient to cough,

Fig. 34.12: External ring invagination test

[1]This test has no meaning or usefulness in bubonocoele. It is a test to differentiate scrotal swelling from inguinoscrotal swelling and assumes significance in complete hernias.

the impulse touches the pulp of the finger in direct hernia and the tip in indirect hernia.

PEARLS OF WISDOM

This test causes discomfort to the patient. It cannot be done in female patients because the labial skin is thick and not lax. Hence, it is not a relevant test. However, in very early doubtful cases of indirect hernia an impulse on cough may be appreciated at the deep ring in this test.

5. **Internal ring occlusion test** (deep ring occlusion test): Reduce the swelling first (Fig. 34.13).

   Locate the deep ring above the midpoint between anterior superior iliac spine and symphysis pubis. Occlude the deep ring with the thumb and ask the patient to cough.

   a. If impulse and the swelling are seen, it is a direct hernia because it occurs in the Hesselbach's triangle (medial to deep ring).
   b. If the swelling is not seen, it is an indirect hernia. Deep ring occlusion test can be done with the patient in standing and supine position.

Problems of deep ring occlusion test

a. If occlusion is not done properly, results may vary.
b. Pantaloon hernia (Romberg hernia, saddlebag hernia, dual hernia). It is a direct hernia having indirect component.

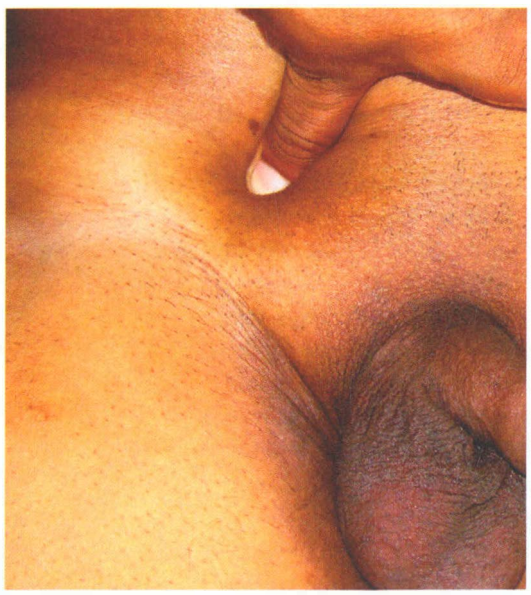


Fig. 34.13: Deep ring occlusion test

6. **Leg raising test** or head raising test (Fig. 34.14)
   - Weakness of oblique muscles is manifested by Malgaigne's bulgings above the medial half of inguinal ligament. ***It is an absolute indication for hernioplasty.***

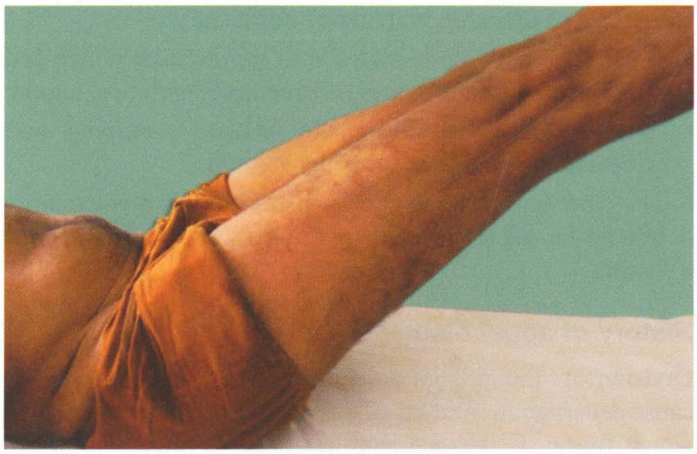


Fig. 34.14: Leg raising test

   - **Malgaigne's bulgings indicate weakness of the oblique muscles of the abdominal wall.**

7. **Zieman's method:** Three finger method

   Keep index finger at deep ring, middle finger on the posterior wall above and lateral to the external ring and ring finger at femoral ring. Now ask the patient to cough. Depending upon the type of hernia, impulse is felt. It is not necessary to perform this test in incomplete or complete indirect hernias.

8. **Per abdomen:** To rule out any mass (colonic).
9. **Look for phimosis/stricture urethra:** Young patients having urinary complaints with hernia may be suffering from stricture urethra. Lift the scrotum and feel for any strictures in the bulbar urethra. Retract skin of prepuce and rule out phimosis.
10. **Per-rectal examination** should be done in elderly patients to rule out prostatic enlargement.
11. **Examination of respiratory system** is done to rule out chronic bronchitis, tuberculosis, etc.

PEARLS OF WISDOM

Examine the opposite side also.

Clinical examination of a hernia in a child

- Swelling may not be visible at first as it may be covered by thick pad of fat. Examine when a child strains (cry), or after child's play (jumping, etc.). Examine the root of scrotum—may find hernial sac (thickening).
- **Gornall's test:** By gentle compression on child's abdomen (hold the child on its back), hernia may become apparent.
- Invagination test is almost impossible. Hence, it is better not to do it.

Diagnosis (one example)

Right side, indirect, incomplete, uncomplicated, reducible omentocoele. Table 34.2 for comparison and Table 34.3 for differences between hernia and hydrocoele.

DIFFERENTIAL DIAGNOSIS OF A GROIN SWELLING

Groin refers to the junction of lower abdomen with the thigh. Hence, swellings in the inguinal region and upper thigh close to the inguinal ligament are included under groin swellings.

1. **Inguinal hernia** (Fig. 34.15)
2. **Femoral hernia:** The main sac is below and lateral to pubic tubercle (Fig. 34.16).
3. **Vaginal hydrocoele:** Fluctuation and transillumination tests are usually positive and getting above swelling is possible. (Please note that in infantile hydrocoele and hydrocoele *en bisac*, getting above swelling is not possible) (Fig. 34.17).
4. **Retractile testis:** It can present as a firm swelling in the inguinal region. Scrotum is empty (Fig. 34.18).
5. **Saphena varix:** Patient can present with a swelling in the thigh. Swelling is usually about 2.5 cm below the pubic tubercle. **A swelling that disappears on elevation of the leg is characteristic of a swelling of venous origin** (Fig. 34.19).
6. **Funiculitis:** A funiculitis can occur with or without acute epididymoorchitis. Severe pain in the inguinal region, tender swelling, high grade fever with chills and rigors are characteristic. Spermatic cord is thickened and swelling is not reducible (Fig. 34.20).
7. **Inguinal lymphadenitis:** Pain and nodular swelling below inguinal ligament is a feature. It is not reducible and some source of infection in the lower limb is usually present (Fig. 34.21).
8. **Lipoma of the cord:** It presents as a soft, lobulated but irreducible swelling in the inguinal region (Figs 34.22 and 34.23).

Investigations

1. Routine investigations such as complete blood picture (CBP) and urine examination are done. In elderly patients, chest X-ray, electrocardiography or even pulmonary function tests may be necessary. Patients with urinary complaints are evaluated for prostatic enlargement and stricture urethra.
2. **Ultrasound:** Hernia is a clinical diagnosis. In the vast majority of the cases, no investigations are required specific to the diagnosis of hernia. However, in appropriate cases imaging can be done.

- In so-called occult cases wherein patient has groin pain but clinically not evident. Ultrasound can detect a sac—however it is operator dependant, ultrasound is also useful in cases of postoperative swelling in the groin to rule out haematoma/seroma/recurrence.

Table 34.2 Differences between direct and indirect hernia

	Direct hernia	Indirect hernia
1. Age	Common in elderly	Can occur in any age group
2. Aetiology	Weakness of posterior wall of inguinal canal	Preformed sac
3. Precipitating factors	Chronic bronchitis, enlarged prostate	—
4. On standing	Pops out	Does not pop out
5. Side	Usually bilateral	Unilateral (30% bilateral)
6. Internal ring occlusion test	Swelling is seen	Swelling is not seen
7. Malgaigne's bulgings	May be present	Absent
8. Complications	Not common because neck is wide	Common, neck is narrow—obstruction and strangulation
9. Relationship of sac to the cord	Sac is posterior to the cord	Sac is anterolateral to the cord
10. Direction of the sac	It comes out of Hesselbach's triangle	Sac comes through the deep ring

Table 34.3 Clinical differences between hernia and hydrocoele

	Indirect complete hernia	Vaginal hydrocoele
1. Standing position	Swelling of scrotum and inguinal region	Swelling confined only to scrotum
2. Impulse on coughing	Present	Absent
3. Getting above swelling	Not possible	Possible
4. Reducibility	Usually present unless complicated	Not reducible
5. Consistency and transillumination	Soft, gurgling, no transillumination	Soft, fluctuant, transillumination is present

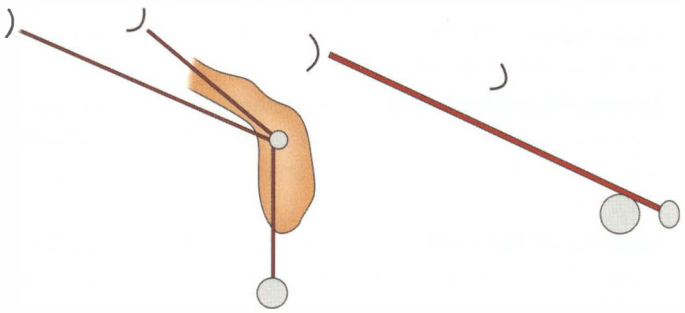


Fig. 34.15: Inguinal hernia

Fig. 34.16: Femoral hernia

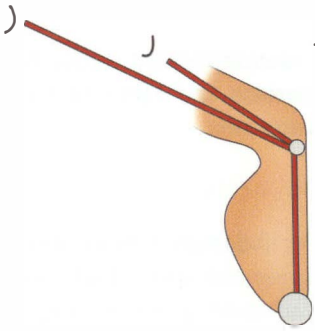


Fig. 34.17: Infantile hydrocoele

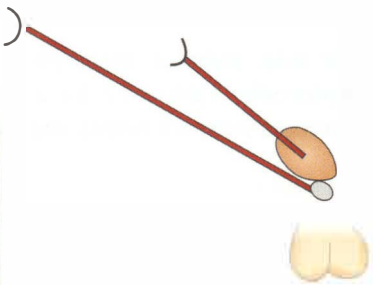


Fig. 34.18: Retractile testis

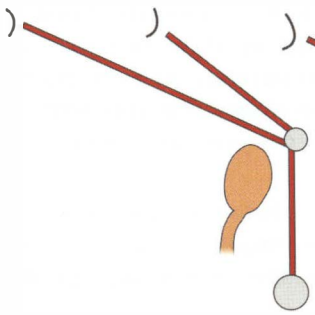


Fig. 34.19: Saphena varix

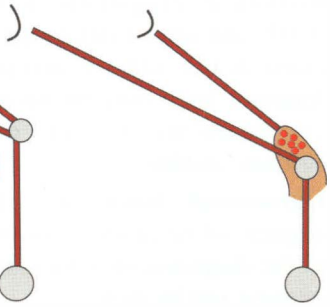


Fig. 34.20: Funiculitis

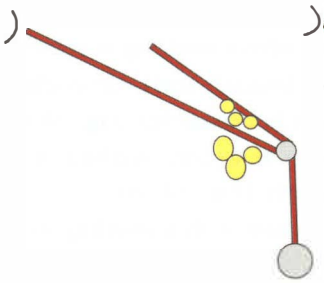


Fig. 34.21: Inguinal lymphadenitis

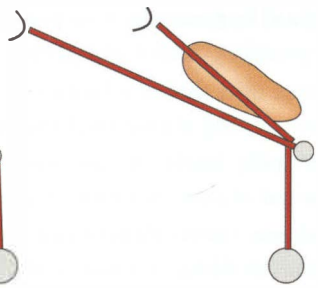


Fig. 34.22: Lipoma of the cord

Fig. 34.23: Left inguinal lipoma of the cord

PEARLS OF WISDOM

Hernia is a clinical diagnosis. If you are asked to give one investigation to confirm hernia (may be in early cases) it is ultrasound.

3. **Computed tomography(CT)** scan is ideal in cases of giant hernias, or special types such as obturator hernia, perineal hernia, etc.
4. **Magnetic resonance imaging (MRI):** Ideal in sportsmen who complain of groin pain, to detect hernia or to rule out muscle sprain or any other orthopaedic disorders.

Preoperative preparation

- A patient with chronic bronchitis and bronchial asthma should be properly treated with bronchodilators, antibiotics, mucolytic agents, etc. Cigarette smoking should be stopped.
- Elderly patients with bilateral hernia mostly suffer from benign prostatic hypertrophy. **Prostatectomy should be considered first followed** by repair of hernia, in such cases. Recent history of constipation and appearance of a hernia should arouse the suspicion of carcinoma colon. Investigate by colonoscopy/fibreoptic sigmoidoscopy before the treatment of hernia.
- Young adults with difficulty in passing urine may have a stricture urethra. They should undergo proper treatment for the stricture. Now it is rare to find a patient with hernia having a stricture urethra.

Treatment

PEARLS OF WISDOM

Herniotomy, herniorrhaphy and hernioplasty are the three "key" operations for inguinal hernia.

1. Herniotomy (Fig. 34.24)

Excision of the sac alone is done in patients up to 14–16 years of age (children). Hernia occurs due to preformed sac. Hence, **no repair is necessary**.

2. Herniorrhaphy

- It can be of two types: Bassini's and Shouldice.
- What is done today is the modified Bassini repair––(Read also original Bassini operation).

A. Modified Bassini's herniorrhaphy (Fig. 34.25).

- Herniotomy with approximation of posterior wall of the inguinal canal by suturing the conjoined tendon (above) to the inguinal ligament below, by using interrupted, nonabsorbable suture material such as nylon, thick silk or polypropylene. This is the most popular method. Repair of stretched deep ring by narrowing and laterally displacing the spermatic cord is done **(Lytle's repair)** in selected cases at the end of the procedure.
- If there is tension, an incision over the anterior rectus sheath will help in doing the repair **(Tanner's slide)**.
- More details are given in Chapter 53 on operative surgery.

Indications for Bassini's herniorrhaphy

- Indirect hernia with good muscle tone.
- Direct hernia with good muscle tone.
- Young patients with good muscle tone.

Fig. 34.24: Herniotomy

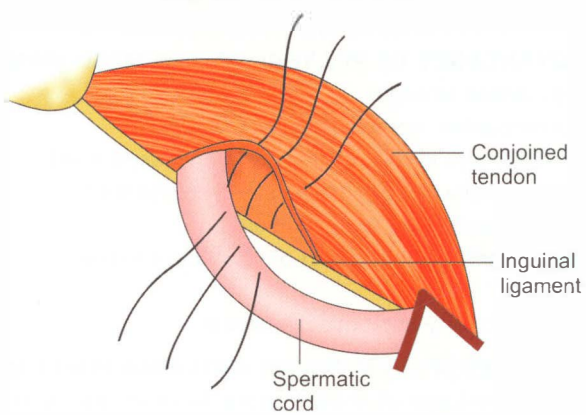


Fig. 34.25: Bassini's herniorrhaphy

Criticism for Bassini's herniorrhaphy

- It is a repair with tension
- Conjoined tendon and inguinal ligament approximation is not physiological.

PEARLS OF WISDOM

Hence irrespective of the type of hernia, mesh repair (Lichtenstein—next page) is recommended today as first line of repair.

What is original Bassini operation?

- Eduardo Bassini incised the external oblique aponeurosis through the external ring.
- Resected the cremasteric muscle
- Divided the transversalis fascia from pubic tubercle to beyond the internal ring.
- Reinforced the posterior wall of the inguinal canal with a single row of interrupted nonabsorbable sutures.
- Sutured the internal oblique muscle, transversus abdominis muscle and upper leaf of the transversalis fascia (triple layer) to the lower leaf of the transversalis fascia and inguinal ligament (double layer) in a single row of interrupted nonabsorbable sutures.
- He then reapproximated the external oblique aponeurosis over the cord structures.
- In modified Bassini repairs, the transversalis fascia is not opened. Cremasteric muscle is not excised. Only conjoined tendon is sutured to the inguinal ligament.

B. Shouldice repair (Key Box 34.5)

- It is the most popular tensionless method wherein only local tissues are used.
- After opening the inguinal canal, herniotomy is done.
- Transversalis fascia which forms the posterior wall, is incised from the internal ring till pubic tubercles.
- Thus, upper and lower flaps of transversalis fascia are sutured in a double-breasting manner by using nonabsorbable sutures such as 34 gauge stainless steel wire, polyamide or polypropylene. This is the **first layer** of Shouldice repair.
- The **second layer** is like Bassini's, where in conjoined tendon is sutured to the inguinal ligament by using nonabsorbable sutures.
- The **third layer** is completed by suturing upper flap of external oblique aponeurosis to the inguinal ligament.
- The results have been good in Shouldice's hands. The operation needs **expertise**.

KEY BOX 34.5

SHOULDICE REPAIR

- **First layer:** Double breasting of the transversalis fascia.
- **Second layer:** Conjoined tendon is sutured to inguinal ligament.
- **Third layer:** Upper half of external oblique aponeurosis is sutured to inguinal ligament.

3. Hernioplasty

Indications

1. Indirect or direct hernia with a good muscle tone. In such cases, darning can be done.
2. Indirect or direct hernia with weak muscle tone, meshplasty is preferred.
3. Recurrent hernia

- Hernioplasty refers to strengthening the posterior wall of inguinal canal. There are two types of hernioplasties which are commonly practised—A. Mesh repair, B. Darning.

A. Strengthening: The posterior wall **(Lichtenstein repair)** (Figs 34.26 and 34.27, Key Boxes 34.6 and 34.7) of inguinal canal by a prolene mesh or Marlex mesh. The fibroblasts and capillaries grow over the mesh, converting it into a thick fibrous sheath and strengthening the posterior wall. The mesh is fixed inferiorly to lacunar and inguinal ligaments, medially to overlap rectus sheath and fixed to fascia over the pubic bone. A few interrupted sutures are put to fix it to the transversalis fascia. **Laterally, an artificial deep ring is created by crossing of both upper and lower leaves of the mesh.** To attain this, a slit is given on one side of mesh. (Lacunar ligament is that portion of the inguinal ligament which extends backwards and upwards to the pectineal line and forms the medial margin of the femoral ring).

Characteristics of the ideal mesh

- Biocompatibility means it should not do any harm, should be chemically and physically inert.
- Risk of infection should not be there
- Handling should be good
- Socioeconomics economical
- Longevity

B. Prolene nylon darning: Suturing the conjoined tendon to the inguinal ligament without tension in a criss-cross manner by using prolene suture material (handmade mesh). This is preferred in direct and indirect hernias (Fig. 34.28) described by Maloney.

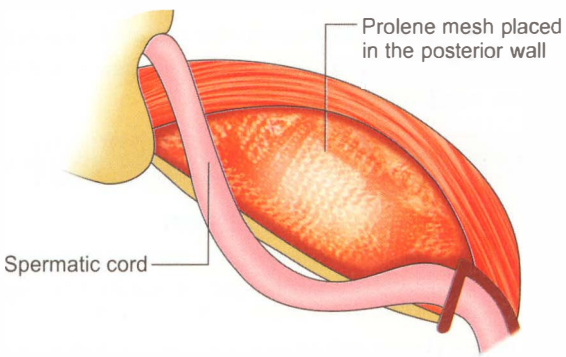


Fig. 34.26: Prolene mesh repair: Lichtenstein repair

KEY BOX 34.6

LICHTENSTEIN REPAIR

- Polyprophylene mesh is used (Fig. 34.27)
- 8 × 16 cm mesh is tailored to patient's requirement.
- **Preparation of mesh:** Corners can be cut so as to give a round shape. A slit is given on the lateral border of the mesh at the junction of lower one-third and upper two-thirds, to allow spermatic cord to pass through. The two tails (slit-ends) are overlapped.
- **Suturing:** Medially, the mesh overlaps the pubic tubercle and is sutured over the tissue of symphysis (avoid pubic bone to prevent osteitis pubis). Laterally, the two tails are placed beyond deep ring and sutured. Inferiorly, it is sutured to inguinal and lacunar ligaments and superiorly to conjoined tendon.

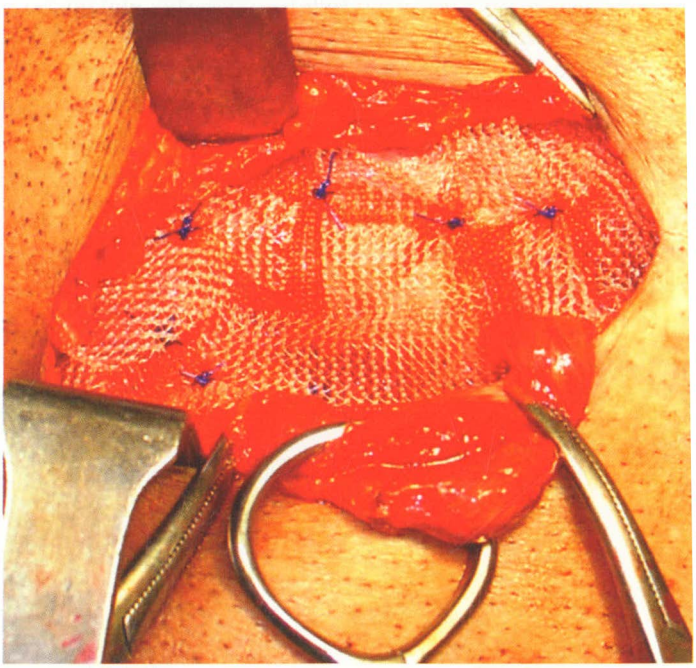


Fig. 34.27: Lichtenstein repair is the most popular type of open hernia repair

KEY BOX 34.7

ADVANTAGES OF POLYPROPYLENE MESH (PPM)

- High tensile strength
- Biocompatible, nonabsorbable
- Monofilament strong, elastic and transparent mesh
- Ideal porosity for high visibility and colonisation
- Strong mechanical reinforcement
- Encourages rapid ingrowth of connective tissue
- Cheaper
- Flexible for any anatomic placement

ADVANTAGES OF LIGHT WEIGHT AND LARGE PORES MESH

- Less shrinkage of mesh, more flexible, better tissue integration, better comfort.

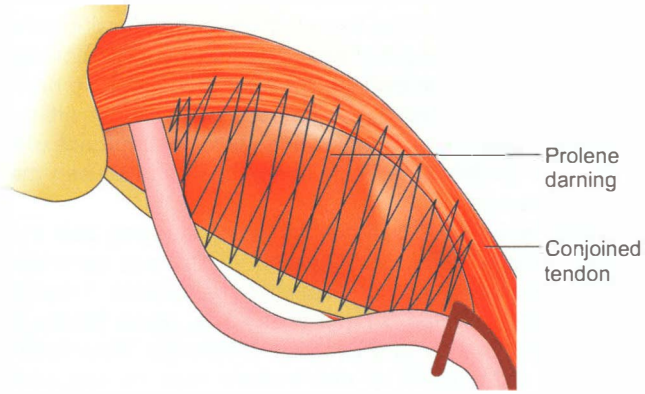


Fig. 34.28: Prolene darning

BIOLOGICAL MESH

These are sterilised sheets of connective tissue derived from human or animal dermis or porcine intestinal submucosa:

- They are decellularised
- Like the mesh, they provide scaffold for connective tissue to grow and collagen deposition.
- Enzymatic reaction takes place in the host, later fibrous tissue formation occur.
- Advantages: Chronic inflammation and foreign body reaction, stiffness and fibrosis, and mesh infection are uncommon—usually do not occur.
- They can be used in presence of infection.
- They are very expensive

Other surgeries for inguinal hernia

1. Kuntz operation (Fig. 34.29)

In this operation the spermatic cord is divided at the deep ring and it is removed along with the testis, so that the deep ring can be permanently closed, and hernia never recurs. It is indicated in elderly patients with recurrent hernia and poor abdominal muscle tone. Hamilton Bailey's operation—cord is divided but testis is retained.

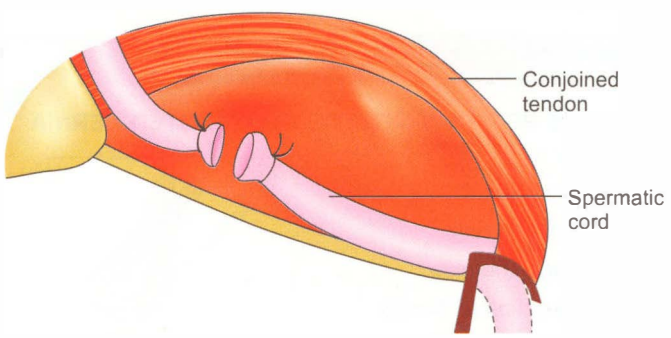


Fig. 34.29: Kuntz operation

2. Andrew's imbrication

In this operation, overlapping of external oblique aponeurosis is done.

3. McVay

It refers to suturing of conjoined tendon to the Cooper's ligament.

4. Nyhus repair

Ideally indicated in bilateral direct hernia or recurrent hernia wherein a broad mesh is kept in the preperitoneal space.

Note: Students should remember that the operation which you have seen in your hospital should be mentioned in the clinical examination.

5. Stoppa repair[1]

The **Stoppa repair is a tension-free type** of hernia repair. It is performed by wrapping the lower part of the parietal peritoneum with prosthetic mesh and placing it at a preperitoneal level over Fruchauds myopectineal orifice. This operation is also known as giant prosthetic reinforcement of the visceral sac (GPRVS).

6. Marcy repair

Simple high ligation of the sac combined with tightening of the internal ring (done in children).

NEW DEVELOPMENTS

1. **What is Dasarda technique?** In this operation, a strip of external oblique aponeurosis is prepared isolated but still connected medially and laterally to the external oblique muscle, and sutured to conjoined tendon and inguinal ligament below.
2. **What is hernia system?** A two layered mesh is used—one to place deep to transversalis fascia (with finger in the deep ring, blind and blunt dissection is done to develop a deep plane) and the other in front of the transversalis fascia.
3. **What is mesh plug repairs?** These are simple plugs of mesh inserted into the deep ring. It is a simple procedure but mesh migration and seroma within the mesh called as Meshoma are common.

COMPLICATIONS OF HERNIA SURGERY

These are common complications after surgery. Often they are mild and note so worrisome. However, some of the complications can be serious which require immediate attention and treatment.

1. Complications during surgery

- **Injury to the iliac vessels:** The most serious but rare complication is injuryto iliac vessels. It can happen in thin patients when suturing of the inguinal ligament is done from

[1]It was first described in 1975 by Rene Stoppa.

lateral to medial side. The sudden jet of fresh red blood indicates that the bite has been taken through the artery. It is better to call the vascular surgeon, extend the incision, have a proximal control, suture directly or do a resection and end to end anastomosis. They have to be anticoagulated with low molecular weight heparin followed by oral anticoagulants.

- **Injury to the urinary bladder:** This can happen when anatomy is not clear as in few giant or scrotal hernias, perineal hernias or distortion due to previous surgery. Sudden finding of clear fluid with urinary smell means bladder injury. Immediate repair with 2–0 vicryl followed by urinary catheter placement for 3 weeks is the treatment.

2. Early postoperative period

- **Pain:** Pain is common due to the incision in the skin and some degree of retraction of structures such as inguinal ligament downwards and conjoint tendon upwards. The pain can be decreased by local anaesthetic infiltration, examples bupivacaine 0.25% up to a maximum of 2 mg/kg body weight.
- **Bleeding:** Perfect haemostasis is the aim of all surgeries. In spite of this, a few bleeders may open up, mostly venous blood—may be pampiniform plexus veins or arterial blood from inferior epigastric artery. The bleeding may stop with compression bandage. Otherwise, exploration and ligation of bleeders needs to be done in the operation theatre.
- **Urinary retention is common, more so in males:** Pain, spinal or epidural anaesthesia, sedatives, lack of privacy are contributing factors. Provide analgesia, privacy and hot fomentation to suprapubic region. If all of these safe, catheterise bladder as a last step.
- **Abdominal distension:** This is not common. It can happen when large intestinal contents of the hernia sac are reduced or handled as in scrotal hernias or sliding hernias. It is also important to realise that omentum is attached to stomach and colon above. One should see that bleeders from injured arteries of the omentum should be ligated properly. Some intraperitoneal blood may add to paralytic ileus.

3. Intermediate—between 3 and 7 days

- **Seroma** is due to inflammatory response to mesh or suture materials. It causes swelling and anxiety that it may be a recurrence. When in doubt, get an ultrasound examination first. Seroma needs to be aspirated. Seroma is more common after laparoscopic hernia repairs.
- **Wound infection:** Hernia is a clean surgery. Infection should not occur. However, poor handling of the tissues, haematoma, seroma and diabetes may precipitate wound infection. Open the sutures, drain the pus and use appropriate antibiotics in such cases. Persistent wound infection may prompt removal of the mesh. A few cases of tuberculosis have been reported. This is due to improper sterilisation of the mesh used.

4. Late:

Late complications are not all that common. One complication which bothers a few patients is chronic pain called inguinodynia.

- **Inguinodynia:** The chief factor is injury to the following nerves: iliohypogastric, ilioinguinal and genital branch of genitofemoral nerve. Injury can be in the form of entrapment of the nerves, traction injury, cauterisation, transection, etc. These are more common after mesh repair because of entrapment of the nerves or perineural fibrosis and adhesions between mesh and the nerves.
  Clinical features include dull aching or dragging pain in the groin, genitalia, suprapubic region. Some may complain of diminished sensation or even hyperaesthesia. Treatment include reassurance, simple analgesics, nerve blocks with anaesthetic agents and injection of steroids. Neurolysis by inguinal exploration or neurectomy may be required in appropriate cases.
- **Testicular atropy:** It is due to injury to the testicular artery which is not noticed during surgery. A few days later, the patient may complain of small testis. Examination reveals no sensation in the testis. Orchidectomy may be required in such cases.

COMPLICATIONS OF HERNIA

1. Irreducibility (Key Box 34.8)

Occurs due to adhesions formed between omentum, sac and the contents. Irreducibility produces dull aching pain.

KEY BOX 34.8

DIAGNOSIS OF IRREDUCIBLE HERNIA

- Hernia is tense
- Tender
- Irreducible
- No impulse on cough
- Recent increase in size of swelling

2. Obstructed hernia (Key Box 34.9 and Fig. 34.30)

Irreducible hernia + obstruction to the lumen of the gut gives rise to obstructed hernia. Clinically, it produces severe colicky abdominal pain, abdominal distension, vomiting and step ladder peristalsis.

KEY BOX 34.9

FACTORS RESPONSIBLE FOR OBSTRUCTED HERNIA

- Narrow neck
- Irreducibility
- Sudden straining
- Too many contents
- Long duration of hernia
- Sliding hernia

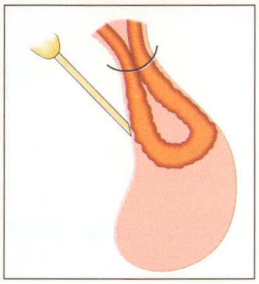


Fig. 34.30: Obstructed hernia

Treatment

Urgent division of the neck of the sac followed by herniorrhaphy or hernioplasty.

3. Strangulated hernia

Irreducibility + obstruction + impairment of blood supply to intestine.

Pathology

- Strangulation commonly occurs in femoral hernia, obturator hernia and in indirect hernia.
- Initially the venous return is occluded, the part gets congested and mucosal ulceration and haemorrhage occurs into the gut wall. It also results in oedema due to capillary exudation.
- If the obstruction is not relieved, constriction of the artery takes place resulting in gangrene of bowel. If this happens, there is a proliferation of bacteria.
- Gangrene appears at the ring of constriction first. Later it develops in the antimesenteric border (Fig. 34.31).
- Such a gangrenous segment contains decomposed blood in which gram-negative organisms multiply. They produce endotoxins resulting in endotoxic shock. If the gangrene extends into the intra-abdominal segment of the bowel, peritonitis can occur.
- The deep ring and the external ring are the common constricting agents.

Clinical features

- Sudden, severe, prolonged pain with some features of shock are indicative of strangulation.
- **Clinical examination of such hernia reveals** (Fig. 34.32):

1. Tense (hernial sac is tense—differentiates it from obstructed hernia)
2. Tender
3. No impulse on cough
4. Irreducible
5. Recent increase in the size of the swelling
6. General condition of the patient is poor:
   - Feeble pulse
   - Hypotension
   - Rebound tenderness
   - Guarding and rigidity, if infection has spread intra-peritoneally.
   - Absent bowel sounds
   - Toxic look

Treatment

I. General measures

- The patient is hospitalised. The foot end of the bed is raised so that an irreducible hernia may reduce by gravity. However, if there is a suspicion of gangrene, this step is not recommended.
- A Ryle's tube is introduced to decompress the stomach, thus preventing vomiting and reducing abdominal distension.
- Intravenous fluids are given to correct dehydration and t prevent renal failure.
- Narcotic analgesics are required to reduce the pain.
- An attempt should be made to reduce the swelling whe there is no gangrene by the following measures:
  A. Good sedation
  B. Patient's thigh is flexed, adducted and medially rotated

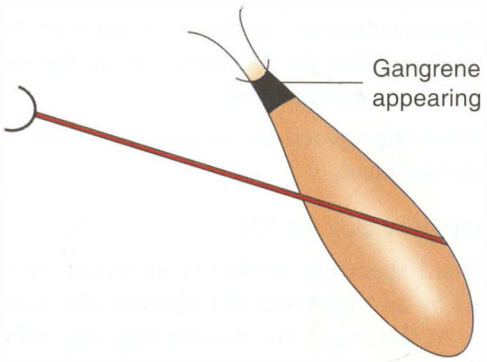


Fig. 34.31: Gangrene appears at ring of constriction first and then progresses

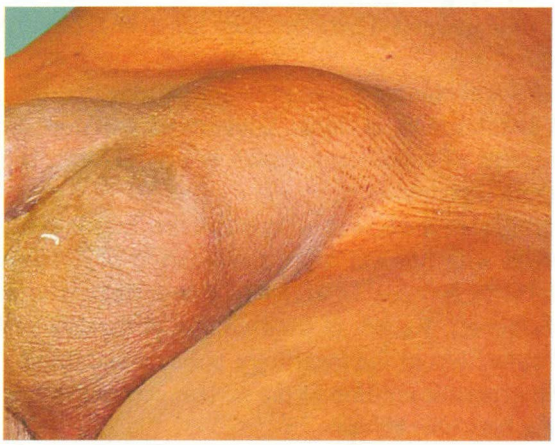


Fig. 34.32A: Strangulated hernia

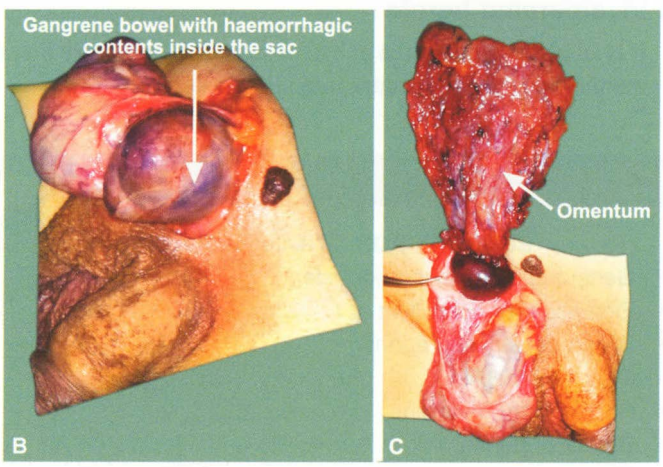


Fig. 34.32B and C: Strangulated hernia with gangrene

C. With the right hand, the sac is gently squeezed by applying pressure over the scrotum. At the same time with the left hand, the proximal portion of the sac is guided into the inguinal canal. This procedure is described as taxis. **Taxis is contraindicated if there is gangrene**.

D. **Complications of forced reduction** include contusion of intestinal wall, rupture of the sac at the neck and reduction *en masse,* i.e. the entire sac with the contents are reduced into the abdominal cavity but the intestine still remains strangulated.

- The patient is prepared for surgery and blood is grouped and cross-matched.

II. Surgery (Key Box 34.10)

- With broad spectrum antibiotic coverage, the hernia is explored by an inguinoscrotal incision and hernial sac is defined. At this stage, ***the constricting ring should not be divided.*** First all the toxic fluid from the sac is aspirated. The constriction is then divided using a grooved director or hernia bistoury. **While dividing the constricting ring, the inferior epigastric vessels which are situated medially may be damaged**. Therefore, care has to be taken to protect these vessels.
- If the bowel is gangrenous, resection of gangrenous segment and anastomosis is done. Closure of incision includes placement of tube drain, which is brought out through a separate incision.
- If viability of the bowel is doubtful, the intestinal loops are covered with hot wet mops for a period of 5–10 minutes and 100% oxygen is given to the patient (request the anaesthetist). Return to pink colour, peristalsis of bowel and pulsations in the mesentery indicate viability.
- If the general condition of the patient permits, repair of the hernia may also be done.
- If evidence of peritonitis is present or if gangrene is spreading within, laparotomy should be done.

4. Incarcerated hernia

It is an obstructed hernia due to obstruction caused by faecal matter. It generally occurs in a sliding hernia

KEY BOX 34.10

SURGERY FOR STRANGULATION

- Generous inguinal incision, identify the sac
- First aspirate toxic fluid
- Divide constricting agent
- Check for viability
- Resection of gangrene
- Repair of hernia
- Broad spectrum antibiotics

5. Inflamed hernia

It occurs when the contents of hernia get inflamed, e.g. appendicitis in a hernial sac, Meckel's diverticulitis in hernial sac.

Thus, complications of hernia can be dangerous (Key Box 34.11). It may range from a simple obstruction to a life-threatening strangulation. Hence, early diagnosis and early treatment are necessary in all cases of indirect hernia. In a few selected cases of direct hernia, wherein the defect is big, the chances of strangulation are less. However, if there are no medical contraindications, surgery has to be advised.

KEY BOX 34.11

SUMMARY OF COMPLICATIONS OF HERNIA

- Irreducibility
- Obstructed hernia
- Strangulation
- Incarcerated hernia
- Inflamed hernia

Following are a few examples of strangulations without obstruction. They have diarrhoea and bleeding per rectum rather than constipation (Key Box 34.12).

KEY BOX 34.12

STRANGULATION WITHOUT OBSTRUCTION

- Omentocoele
- Richter's hernia
- Littre's hernia

RECURRENT HERNIA

Incidence of 10% recurrence are common medially.

Causes

I. Preoperative

1. Chronic cough
2. Weak muscle tone
3. Straining while passing urine, constipation
4. Obesity, ascites, anaemia.

II. Intraoperative

1. **Improper excision of the sac:** The sac should be ligated at the level of deep ring (neck). This is called **high ligation of the sac**. Very often, the sac is seen as soon as the inguinal canal is opened. If it is ligated at the fundus or body (low ligation), it invariably results in recurrence. A missed indirect sac can be a cause of recurrence (Figs 34.33 and 34.34A and B).

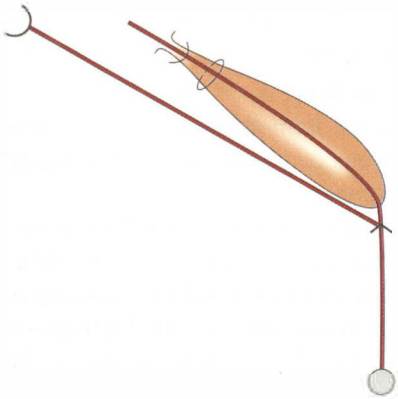


Fig. 34.33: Indirect incomplete hernia

Fig. 34.34A: High ligation

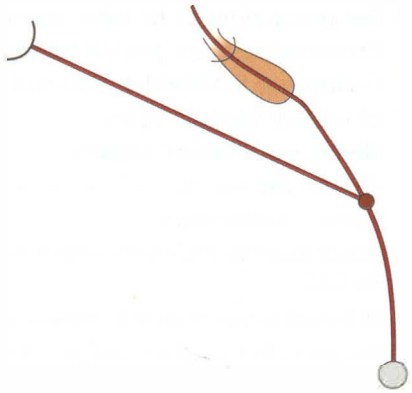


Fig. 34.34B: Low ligation—cause for recurrence

2. **Absorbable sutures** such as catgut have lifespan of 2–3 weeks. If they are used for reconstruction, they invariably result in recurrence.
3. **Bleeding:** At the end of the surgery, small bleeding points should be coagulated by using diathermy or ligatures. Haematoma formation predisposes to infection, which can be the cause of recurrence.
4. **Tension** between suture lines can cause strangulation and fibrosis of muscle fibres. Hence, care and gentleness is important while suturing conjoined tendon to the inguinal ligament.

III. Postoperative

1. **Persistent postoperative cough** weakens the suture line.
2. **Haematoma** can get secondarily infected resulting in pus formation. The sutures give way leading to recurrence. Hence, if there is a significant haematoma, it should be drained.
3. **Infection:** Even though hernia is a clean surgery, chances of infection are present specially in diabetics, alcoholics and immunocompromised patients. Prophylactic antibiotics such as 2nd generation cephalosporin should be given. If infection occurs, it should be treated accordingly.
4. **Exertion:** Too much exertion in the postoperative period, in the form of lifting heavy weights or carrying heavy weights on the shoulder, may weaken the suture line, resulting in hernia.

Most of the recurrences occur within one year. Incidence of recurrent hernia may vary from 2 to 8% even in experienced hands. In a case of recurrent hernia, it is difficult to say whether it is a direct hernia or indirect hernia. From the management point of view, it does not matter.

Treatment

- If the sac is present due to incomplete excision at the previous surgery, it should be completely excised up to the level of deep ring, followed by hernioplasty.
- Meshplasty is the surgical treatment for a recurrent hernia. However, if mesh cannot be placed either due to infection or due to nonavailability, prolene darning can be done.
- Tuberculosis must be ruled out in cases of persisting infection.
- In all these cases, precipitating factors if any, should be treated first.
- The only way to totally prevent recurrence is by closure of the deep ring. This can be done only after dividing the spermatic cord (Kuntz procedure). It is indicated in elderly patients who have multiple recurrences.

SPECIAL HERNIAS

1. Giant hernia: Sac extends up to mid-thigh (Fig. 34.35)

Definition: A giant inguinoscrotal hernia is defined as a hernia that extends below the midpoint of the inner thigh in the standing position.

Clinical features

- Most patients would have had their hernia for several years.
- The contents often include colon, small intestines and bladder.
- A few of these are also sliding inguinal hernias.
- Hence, more prone for complications such as incarceration, intestinal obstruction and scrotal ulceration. The last complication is due to pressure necrosis or due to friction while walking or moving.
- Differential diagnosis includes scrotal elephantiasis.

Fig. 34.35: Giant hernia

- A few precautions to be taken during surgery are:
  - Preoperative chest physiotherapy.
  - Catheterise the bladder. This will decrease the incidence of urinary bladder injury.
  - Bowel wash before surgery.
  - Dissect the sac carefully all around.
  - Perfect haemostasis.
  - Omentectomy and rarely colectomy may be required (Fig. 34.36).
  - If bowel is not resected, mesh can be placed safely.
  - In cases of emergency and patient with co-morbid factors including cardiac problems, take consent for orchidectomy—the best option will be to divide the cord at the level of deep ring and close the deep ring during orchidectomy.

Fig. 34.36: Excised omentum in the kidney tray

2. Dual hernia: It has two sacs, one direct and another indirect, connected by an isthmus which is behind the inferior epigastric artery. It is also known as saddlebag hernia, **pantaloon hernia**, dual hernia or Romberg hernia.

Significance

- Deep ring occlusion test: The inference of the test may not be correct.
- It is the cause of recurrence if one sac is not treated properly.

3. Prevesical hernia: It is also called **funicular direct hernia**. It is a hernia containing portion of the bladder with prevesical fat through the defect in the conjoined tendon on the medial side. History of the swelling becoming less prominent after micturition may be present. Due to narrow neck, it is prone for strangulation.

4. Littre's hernia (Fig. 34.37): It is referred to a hernia containing Meckel's diverticulum. When the diverticulum gets infected, such hernias are called inflamed hernias. The cause of infection may be precipitated by partial obstruction to the diverticulum by constricting agents.

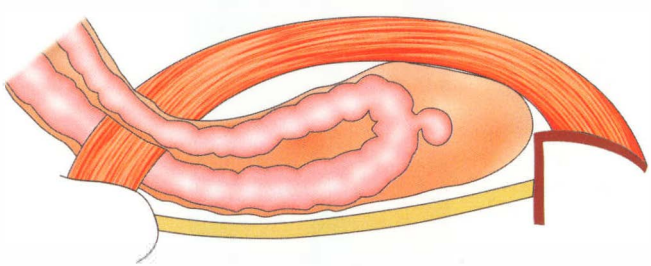


Fig. 34.37: Littre's hernia

5. Maydl's hernia (Hernia-en-W) (Fig. 34.38)

- It is a **W** hernia wherein the intra-abdominal bowel loop segment becomes gangrenous very early but in the scrotum there are no signs of gangrene.
- The patient presents with obstructed hernia and at operation the inguinoscrotal segment has no gangrene. The intra-abdominal segment must be examined and the gangrenous portion should be excised. It can also be called retrograde strangulation. Clinically, there is tenderness above the inguinal ligament.

6. Richter's hernia (Fig. 34.39)

- When only part of circumference of bowel becomes strangulated, it is called Richter's hernia.
- It may spontaneously reduce
- Thus, gangrene may be overlooked at operation
- Even though the patient has features of intestinal obstruction, there will be diarrhoea and often blood in stools.

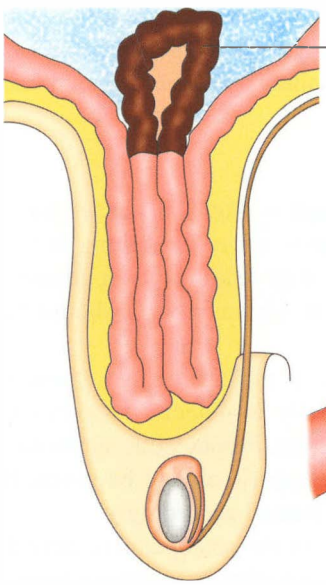


Fig. 34.38: Maydl's hernia

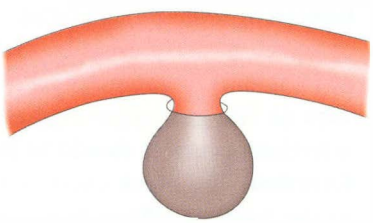


Fig. 34.39: Richter's hernia

- Femoral hernia, obturator hernia are a few examples of hernia which can sometimes present as **Richter's hernia**.

7. Sliding hernia (Hernia-en-glissade) (Figs 34.40A to C)

- Incidence: 1–3%
- Always acquired hernia
- It occurs as a result of the slipping of posterior peritoneum along with the retroperitoneal viscus. As a result of which, the caecum, on the right side and the sigmoid colon on the left side, form the posterior wall of the sac.
- **If the caecum and appendix are the contents of the hernial sac, it is not a sliding hernia.**

- However, a true hernial sac containing omentum or intestines exists.
- Weakness of the abdominal wall at the deep ring lateral to inferior epigastric vessels is a contributing factor also.
- Urinary bladder can also be the content of the hernial sac (Fig. 34.40A).

Clinical features

- It almost always occurs in males
- It commonly affects elderly patients
- It can be suspected when there is a large hernia descending down into the scrotum.
- Left-sided is more common than right-sided hernia.
- It practically always occurs in long-standing cases of inguinal hernia.
- They are not completely reducible.
- It can be direct hernia and indirect hernia also.

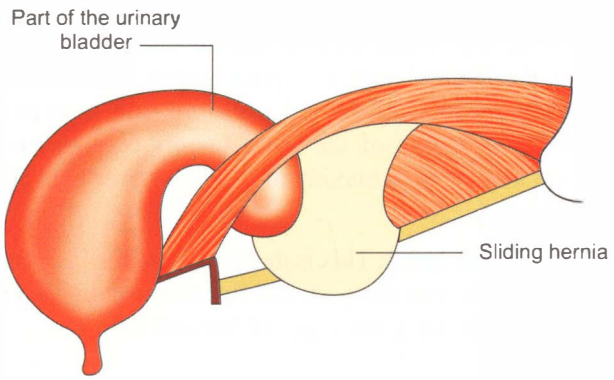


Fig. 34.40A: Sliding hernia

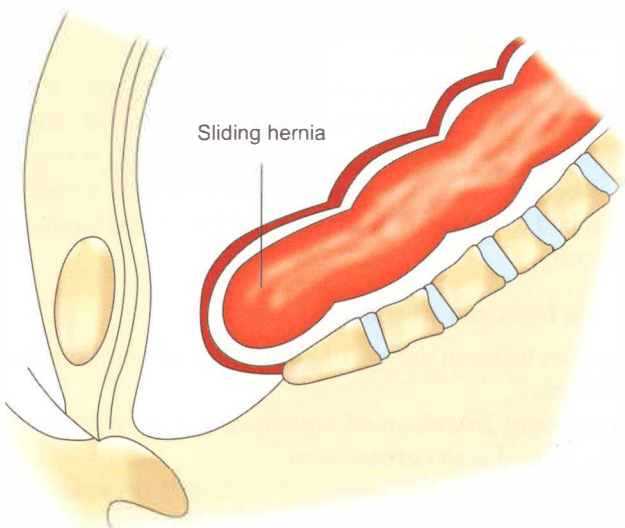


Fig. 34.40B: Sliding hernia

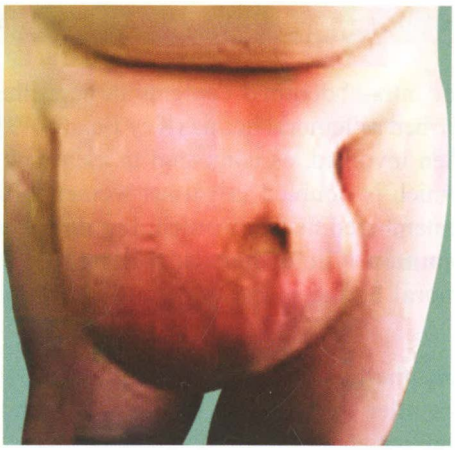


Fig. 34.40C: Sliding hernia (*Courtesy:* Vladimir M Lobankov, Gomel State Medical University, Belarus, USSR)

Complications

These hernias can easily strangulate and since its wall contents include the large intestine, the mortality and morbidity increases.

Treatment

- Truss is absolutely contraindicated.
- Once the hernial sac is opened, the sac **should not be twisted**.
- A purse string suture is applied within to avoid injury to caecum/sigmoid colon.
- The sac is removed and the repair of the hernia done.
- In elderly patients, orchidectomy is advised to give a permanent cure for hernia.

Sliding hernia —Key points

- Always acquired
- Always (almost) in males
- Always occur in long standing hernias
- Always irreducible
- Always colon or urinary bladder forms the wall of the sac
- Always requires a repair
- Always technical difficulties are encountered by junior surgeons while operating on these patients

8. Sportsman's hernia

- This is common in men who play rugby or football wherein injury due to the ball may occur.
- Pain is in the groin radiating to scrotum and upper thigh.
- On examination, there may be tenderness in the inguinal region.
- Orthopaedic disorders must be ruled out first by MRI or CT scan. They are soft tissue injury in the groin, pubic bone diastasis, adductor spasm, etc.
- If hernia is due to tearing of muscles **(Gilmore's groin)**— it should be repaired in the usual manner.

FEMORAL HERNIA

Herniation of intra-abdominal contents through the femoral canal is described as femoral hernia (Key Box 34.13). Women are more often involved, as compared to men with the ratio being 2:1, which is doubled in parous women. However, it should be remembered that **in women, inguinal hernias are the most common type of hernia, followed by incisional hernia.** Femoral hernia is the third most common type of hernia.

Commonly the hernia is unilateral, the right side being affected more often than the left side. It is bilateral in about 15–20% of the patients.

Anatomy of femoral canal and femoral ring
(Fig. 34.41)

- The femoral canal extends from the femoral ring to the saphenous ring. It is 1½ inches below and lateral to the pubic tubercle. It is the innermost compartment of femoral sheath.
- It is similar to a truncated cone **which is narrow at the femoral ring**.
- **Contents of femoral canal are**
  - Fat
  - Fascia
  - Lymphatics: Lymph node of Cloquet
- Femoral vein is in the middle compartment of the femoral sheath and femoral artery is in the lateral compartment.
- Femoral nerve is outside the femoral sheath.
- Femoral sheath: Fascia transversalis is continued downwards behind the inguinal ligament as the anterior layer of the femoral sheath. Fascia iliaca continues behind the femoral vessels as the posterior layer of the femoral sheath.

Boundaries of femoral ring

- ***Anterior:*** Inguinal ligament
- ***Posterior:*** Ligament of Cooper, iliopectineal ligament.
- ***Medial:*** Lacunar ligament (Gimbernat's ligament)
- ***Lateral:*** Thin septum which separates the femoral canal from femoral vein (silver fascia).

KEY BOX 34.13

FEMORAL HERNIA: COVERINGS

- Sac
- Fat and lymphoid tissue
- Transversalis fascia
- Cribriform fascia
- Superficial fascia
- Skin

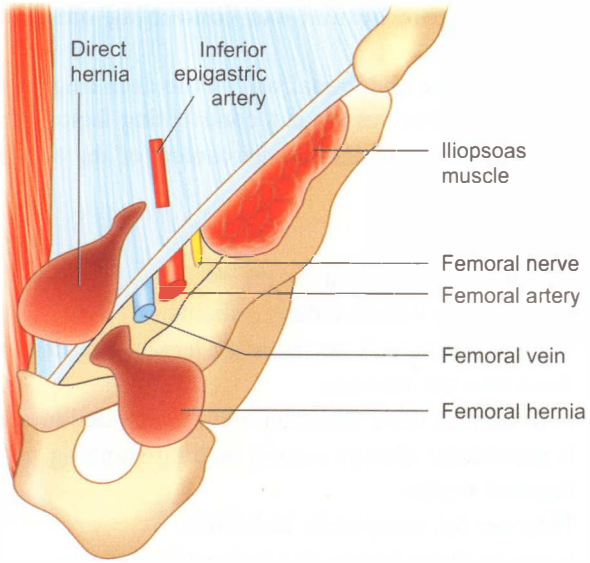


Fig. 34.41: Femoral hernia: Anatomy

Causes for femoral hernia

1. **Pregnancy:** As the gravid uterus compresses the external iliac vein, the empty femoral sheath on the medial side allows the femoral vein to expand within femoral sheath. Thus, increased abdominal pressure due to repeated pregnancies is one of the chief factors responsible for femoral hernias. The maximum incidence is around 30–40 years of age.
2. **Wide femoral canal:** This is due to the narrow insertion of iliopubic tract into the pectineal line of the pubis and may be responsible for a few cases of femoral hernia.

PEARLS OF WISDOM

Femoral hernia is never congenital.

Course of the hernial sac

- As the hernia comes into the femoral canal, it is an oblong swelling due to the rigid femoral canal. When it comes out through the saphenous opening, it expands and becomes retort shaped because Scarpa's fascia is attached to the deep fascia of thigh below the saphenous opening.

Clinical features of femoral hernia

- Females between the age of 20 and 40 years are commonly affected.
- **Gaur sign:** Dilatation of superficial epigastric/circumflex iliac veins due to compression.
- Right side is more commonly affected because of the dominant nature of right side of the body.
- To start with, there is a small swelling below the inguinal ligament, which goes unnoticed very often.

- Expansile impulse is often not present due to the narrow canal.
- Reducibility may be present
- Typically, the swelling is below and lateral to the pubic tubercle (inguinal hernia is above and medial to pubic tubercle) (Fig. 34.42).
- Many (30–80%) present with strangulation.

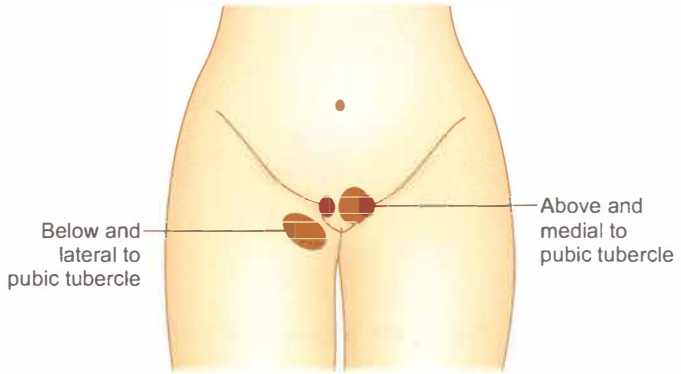


Fig. 34.42: Femoral hernia (R) and indirect hernia (L)

Treatment (Key Box 34.14)

1. Low operation of Lockwood

Incision is placed directly over the swelling in the thigh. The sac is carefully dissected out without damaging the femoral vein. The sac is ligated at the neck, excised and the hernia is repaired—the **inguinal ligament is sutured to Cooper's ligament** (iliopectineal ligament) thus obliterating the femoral ring. Nonabsorbable suture such as prolene or ethilon is ideal. Low approach is indicated in uncomplicated hernia. It is very difficult to manage a gangrenous loop of bowel with this approach.

2. Inguinal operation

- Through an **inguinal incision,** the inguinal canal is opened. The transversalis fascia is incised. Hernial sac is visualised. This is followed by excision of the sac. The high approach is preferred when there is a strangulated femoral hernia. This offers a very good view of the abnormal obturator artery from above, if it is present.
- Repair is done by suturing the conjoined tendon to iliopectineal line.

3. Combined approach: High operation of McEvedy

Inguinofemoral approach: A vertical incision is made over the swelling and extended above the inguinal ligament, and the sac can be dissected from both above and below (look for abnormal obturator artery—see below). This approach has the advantages of both operations mentioned above.

4. Henry's approach

Lower midline for bilateral hernia.

Abnormal obturator artery

Normal obturator artery is a branch of the internal iliac artery. It gives a pubic branch which anastomoses with pubic branch of the inferior epigastric artery. Occasionally, this anastomosis is large and obturator artery then appears to be a branch of the inferior epigastric. Usually it passes lateral to the femoral canal in contact with the femoral vein. Occasionally, the abnormal artery may lie along the medial margin of the femoral ring, i.e. along the free margin of the lacunar ligament. This artery is in danger during surgery for obstructed femoral hernia.

Complications of femoral hernia

1. As the femoral ring and the neck of the sac are narrow, obstruction and strangulation are very common.
2. Richter's hernia
   - Commonly seen in femoral hernias and obturator hernias, which have narrow necks.
   - This occurs when a **portion of the circumference of the bowel** is caught within the hernial sac and which is constricted by the narrow ring. Signs and symptoms of intestinal obstruction are absent, even though it is an obstructed hernia, because the lumen is not obstructed.
   - The hernia is tense, tender, irreducible and has no cough impulse.
   - As the lumen is patent, there may be bloody diarrhoea rather than constipation. Gangrene can occur soon.
   - **Treatment:** Combined or inguinal approach to deal with gangrene.

Summary of femoral hernia (Key Box 34.15)

KEY BOX 34.14

SURGERY OF FEMORAL HERNIA

- Should be done as early as possible, once the diagnosis is made.
- For elective repair, low (femoral) approach: Incision directly over the swelling is ideal. Injury to abdominal obturator artery can occur in this route (found on lateral side in 20% of cases)
- **Transinguinal (Lothiessen):** Can use it when there is gangrene. However, it may weaken inguinal canal.
- Combined: High approach is the choice for strangulated femoral hernias. Approximate inguinal ligament and pectineal ligament.

KEY BOX 34.15

FEMORAL HERNIA

- Rarely occurs in males (5–10%)
- Commonly associated with Richter's hernia
- Fatty female with small swelling under a big belly usually goes undetected.
- Dangerous because of early strangulation
- Cannot be controlled by a truss
- Surgical repair is a must

RARE TYPES OF FEMORAL HERNIA

1. **Lacunar hernia (Laugier's hernia)**
   In this case, the hernia passes through a small defect in the lacunar ligament.
2. **Prevascular hernia**
   In this case, the hernial sac is located behind the femoral vessels and the inguinal ligament. It may be associated with congenital dislocation of the hip (Narath's hernia).
3. **Pectineal hernia**
   In this case, the hernia passes between the pectineus muscle and its fascia, behind the femoral vessels. It is also called Cloquet's hernia.
4. **External femoral hernia**
   It is a hernia lateral to the femoral artery (Hesselbach's hernia).
5. **Narath's femoral hernia**
   In congenital dislocation of the hip, femoral hernia occurs behind femoral vessels. Hence, femoral artery pulsations will be very prominent.

DIFFERENTIAL DIAGNOSIS OF FEMORAL HERNIA

1. **Inguinal hernia:** An inguinal hernia is above and medial to pubic tubercle. The femoral hernia is below and lateral to pubic tubercle (Fig. 34.43).
2. **Saphena varix:** It is the dilated, saccular, upper end of long saphenous vein with varicosity. It disappears on lying down because of gravity. Thrill may be felt on coughing (Fig. 34.44).
3. **Lipoma:** Soft and lobular, slips under palpating fingers (Fig. 34.45).
4. **Femoral artery aneurysm** is rare. It presents as a pulsatile swelling in the groin with a continuous murmur. Peripheral pulses are often weak (Fig. 34.46).
5. **Enlarged femoral lymph nodes** are firm and round. They can be enlarged in lower limb infections, abrasions, wounds in the perineum and also in carcinoma penis (Fig. 34.47).
6. **Psoas bursa:** Osteoarthritis of the hip can produce distension of psoas bursa, which disappears on flexing the hip. Tuberculosis spine can present as iliopsoas abscess (Fig. 34.48).

Fig. 34.43: Large direct hernias

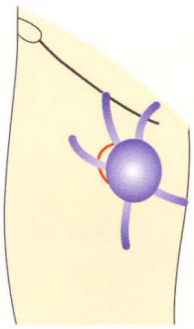


Fig. 34.44: Saphena varix

Fig. 34.45: Left inguinal lipoma of the cord

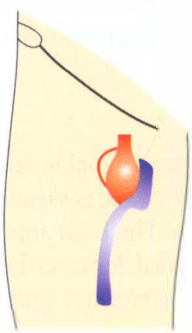


Fig. 34.46: Femoral artery aneurysm

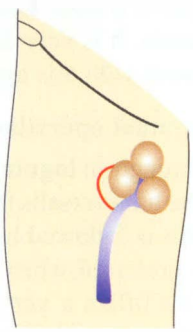


Fig. 34.47: Lymphadenitis

Fig. 34.48: Psoas bursa

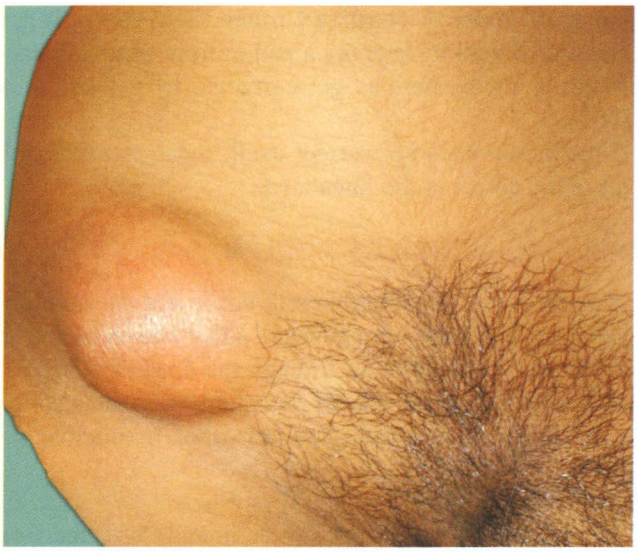


Fig. 34.49: Iliopsoas abscess

7. **Psoas abscess:** It is an iliopsoas abscess due to tuberculosis of spine. There are two swellings, one above and one below the inguinal ligament. Cross fluctuation can be elicited between these two swellings. Tenderness over the spine and X-ray of the spine help in arriving at a diagnosis (Fig. 34.49).

UMBILICAL HERNIA

It can be discussed under three headings:

I. Umbilical hernia of newborn
II. Umbilical hernia of infants and children
III. Umbilical hernia of adults.

UMBILICAL HERNIA OF NEWBORN

- It is called *Omphalocoele—Exomphalos*.
- It is found 1 in 6000 live births.
- Failure of midgut as a **whole or part** to return into coelomic cavity during embryonic life results in exomphalos.
- It is also associated with weakness of abdominal musculature (few fibres may be absent). Two types have been recognised.

1. Exomphalos minor (Fig. 34.50A)

- In this condition, the umbilical cord is attached to the summit of the sac.
- Sac is small and defect less than 5 cm.
- It is treated by twisting the cord and ligating the sac. Care should be taken to avoid damage to the intestine. For example, nursing the child preoperatively in prone position can damage intestines.

2. Exomphalos major (Fig. 34.50B)

- In this condition, the umbilical cord is attached to the inferio aspect of the sac, containing intestines, abdominal structures e.g. liver, bowel.
- Many children are still-born.
- This type of hernia is usually associated with absen abdominal musculature.
- The operation should be done before the rupture of the sac as the morbidity increases greatly in the event of a rupture of the sac.
- During the operation, skin flaps are raised on both sides to cover the defect. A true repair is necessary and is done at a later date.
- *See* also Figs 34.51 and 34.52.

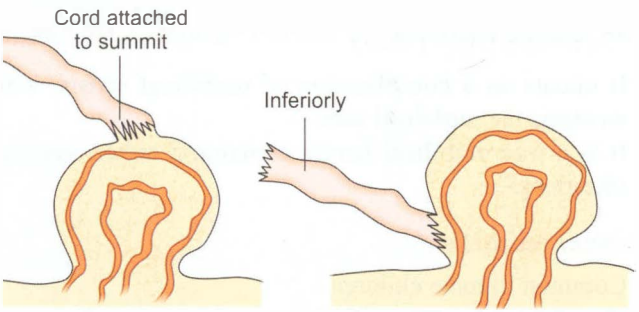


Fig. 34.50A: Exomphalos minor **Fig. 34.50B:** Exomphalos major

Fig. 34.51: Epigastric heterophagus with exomphalos major

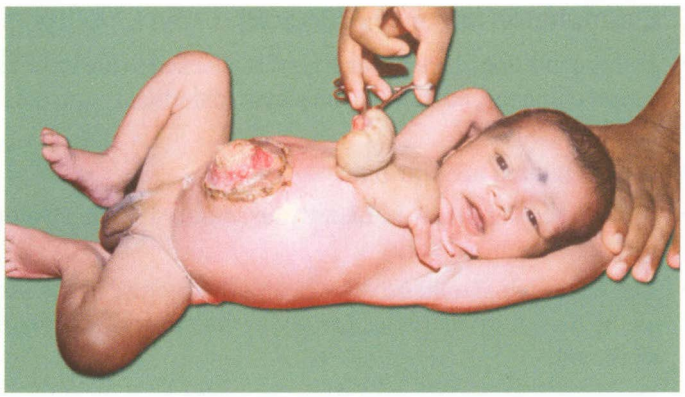


Fig. 34.52: Same child on the 21st postoperative day. The repair is done by rotating flaps from gluteal region of the parasitic baby (*Courtesy:* Prof Vidyadhar Kinhal, Late Prof RS Channagiri, Dr Channanna, Department of Surgery, Vijayanagar Institute of Medical Sciences, Bellary, Karnataka)

UMBILICAL HERNIA OF INFANTS AND CHILDREN

- It occurs as a complication of umbilical sepsis, which weakens the umbilical scar.
- It is a true umbilical hernia containing either omentum/intestines.

Clinical features

- Common in male children
- The child is brought with the complaint of swelling in the umbilical region whenever the child cries.
- Most of the cases are symptomless. Parents are anxious about the swelling.
- Strangulation is rare (Fig. 34.53)

Treatment

- **Reassurance** is the most important advice given to the parents.
- **No treatment is required** other than **strapping** the abdominal wall by keeping a pad in front of umbilicus.
- Majority of the hernias get corrected by 2 years of age (90%).
- If the hernia does not correct itself, repair is necessary to close the defect in the linea alba.

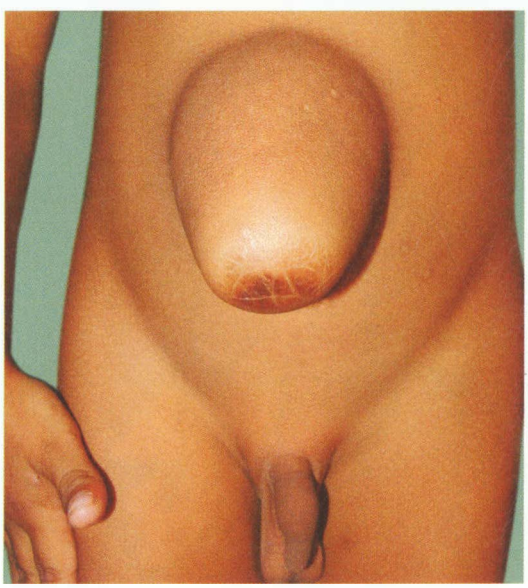


Fig. 34.53: Umbilical hernia in a child

UMBILICAL HERNIA OF ADULTS

- It is not a true umbilical hernia but it is a para-umbilical hernia in which the hernia occurs either above, below or to the side of the umbilicus, through the linea alba.
- The contents are the greater omentum, transverse colon or small bowel. Due to adhesion, it is often irreducible.

Aetiology

- Females in the 5th decade are commonly affected. Male: female ratio is 1:5.
- **Obesity** with flabby abdominal muscle predisposes to para-umbilical hernia.
- Repeated **pregnancies** also weaken the abdominal wall.
- Ascites may precipitate hernia specially in cirrhotic patients.

Clinical features

- Patient presents with a swelling in the umbilical region, which increases on straining or coughing.
- On asking the patient to cough, **expansile impulse** is present (Fig. 34.54).
- They may also have inguinal hernia.
- **Reducibility can be present**
- **Dragging pain** is usually due to omentum which is felt as a firm or granular mass. If gurgling is present, it is indicative of small intestines in the hernial sac.

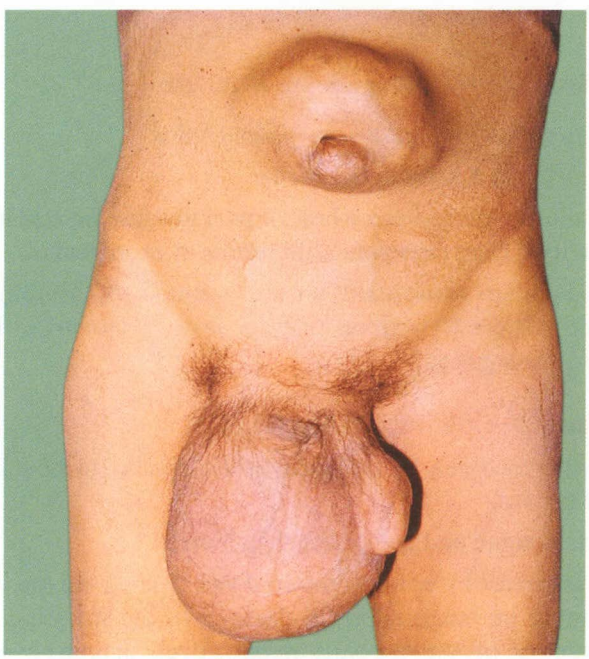


Fig. 34.54: Large direct hernia with umbilical hernia

Table 34.4 Comparison between umbilical hernia in infants and adults

Features	Infants	Adults
Age in years	0–3	50–60
Sex	Common in male child	Common in females
Causes	Neonatal sepsis	Obesity, weak muscles, pregnancy
Defect	A small defect in the umbilical scar	Above or below the umbilicus
Symptoms	Symptom less	Symptoms are present
Strangulation	Rare	Very common
Treatment	Conservative (strapping), surgery (rare)	Mayo's repair

- After reducing the swelling, the **defect** can be made out in the linea alba.
- *See* Table 34.4 for comparison between umbilical hernia in infants and adults.

Complications

1. **Irreducibility** is common due to adhesions between omentum and the sac.
2. **Obstruction** presents with colicky abdominal pain and vomiting. Distension follows soon. Untreated cases develop strangulation. Very often, these patients present with **incarcerated** hernia due to the presence of transverse colon in the sac. They require urgent intervention, failing which gangrene will set in.
3. As the sac enlarges, due to its weight and gravity, it sags down resulting in friction of the skin and this causes **intertrigo** (Fig. 34.55).

Treatment

1. Reduction of weight
2. **Anatomical repair:** Small defects can be closed with non-absorbable sutures such as nylon or prolene (Fig. 34.56).

Fig. 34.55: Umbilical hernia in a cirrhotic patient with skin changes and ulceration

Fig. 34.56: True congenital umbilical hernia in an adult can be repaired by simple closure of the defect by nonabsorbable sutures

3. **Most favoured** surgery for umbilical hernia is **mesh repair**. It is a tensionless repair. It can also be done by **laparoscopic method** which is popular today.
4. ***Mayo's repair*** (surgical treatment, Fig. 34.57 and Key Box 34.16)

- A curvilinear incision is made below the umbilicus or a double semilunar incision can be used. Umbilical cicatrix is then removed (in small hernias it can be preserved).
- Skin flaps are raised (upper and lower)

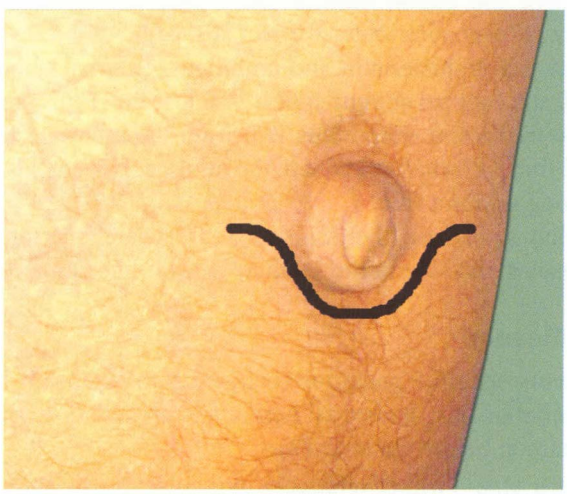


Fig. 34.57: Curvilinear incision

KEY BOX 34.16

MAYO'S UMBILICAL HERNIORRHAPHY

- Excision of umbilicus
- Reduction of contents and excision of the sac
- Double breasting of the fibrous aponeurotic layer
- Haemostasis, suction and obliteration of dead space
- Additional lipectomy and umbilicoplasty

- The sac is dissected all around and the defect in the linea alba is defined.
- Contents of the sac can be reduced with or without opening of the sac.
- Extra **redundant** sac is excised
- Peritoneum is closed
- Defect or cut in the linea alba is extended on both sides **(laterally)** and then upper and lower aponeurotic flaps are sutured together by using double breasting technique.
- Suture material used is prolene or nylon.

INCISIONAL HERNIA

It is also called **ventral hernia** or postoperative hernia. It is a hernia that occurs through a weak scar. Very common in females.

ABDOMINAL WALL — SURGICAL ANATOMY

Contents

It consists of **skin, muscles, aponeurosis, linea alba, sheaths, ligaments, openings—rings**, blood vessels and nerves. Anatomically weak areas are the rings, junctions, empty spaces and where blood vessels pierce abdominal wall.

Formation of rectus sheath (Fig. 34.58)

- Above the costal margin, only external oblique with aponeurosis contributes for rectus sheath.
- Between xiphisternum and umbilicus, external oblique is in front. Internal oblique splits to enclose the rectus muscles. The transverse abdominus is behind the internal oblique. All fuse to form linea alba in the midline. Hence, this is the strong midline area.
- Below semilunar line: All 3 aponeurosis are anterior to the muscles and fuse in the midline to form linea alba.

Significance

- Rectus sheath—posterior rectus sheath is absent below semilunar line. Incisional hernia and spigelian hernia are common below the umbilicus.
- Linea alba—white, relatively avascular, broad above and narrow below. It is the strongest layer of the abdominal wall. Hence, during the closure of the midline incisions, it is important to include good bites through linea alba.
- Umbilicus—strong fibrous ring. Umbilical hernias are common in children due to childhood umbilical infections, in obese patients due to weak muscles and in multiparous woman due to stretching of the muscles due to repeated pregnancies.

Muscles of the anterolateral abdominal wall—oblique muscles

- The anterolateral abdominal wall is made up mainly of muscles. On either side of the midline there are four large muscles. These are the *external oblique*, the *internal oblique*, the *transversus abdominis* and the *rectus abdominis*. Two small muscles, the *cremaster* and the *pyramidalis* are also present. The external oblique, the internal oblique and the transversus abdominis are large flat muscles placed in the anterolateral part of the abdominal wall. Each of them ends in an extensive aponeurosis that reaches the midline. Here the aponeuroses of the right and left sides decussate to form a median band called *linea alba*. The rectus abdominis runs vertically on either side of the linea alba. It is enclosed in a *sheath* formed by the aponeuroses of the flat muscles named above.
- Tendinous intersections—rectus sheath haematoma will be confined within the sheaths as it is prevented from spreading due to tendinous intersections.

Precautions during surgery

- Nerve supply—lower six thoracic and first lumbar nerves. They enter the rectus sheath laterally. In cases of paramedian incisions, after opening the anterior rectus sheath, rectus should be retracted laterally to define and incise the posterior rectus sheath.
- Blood vessels run within the rectus sheath. Rupture of inferior epigastric artery is a known entity resulting in a haematoma below the umbilicus (*see* page 871). Differential diagnosis includes spigelian hernia.
- Weak muscles—interstitial hernias—prune-belly syndrome: A partial or complete lack of abdominal muscles. There may be wrinkly folds of skin covering the abdomen. An undescended testicles in males.

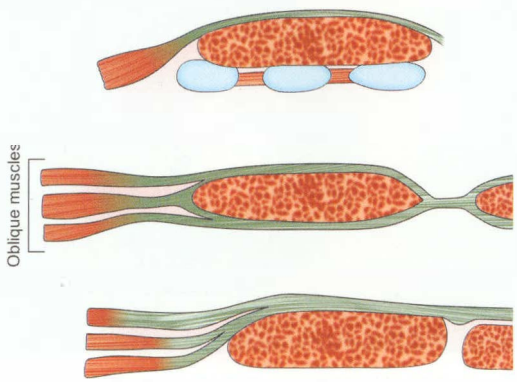


Fig. 34.58: Rectus sheath formation

FACTORS WHICH PRECIPITATE INCISIONAL HERNIA

(Key Box 34.17)

KEY BOX 34.17

CAUSES OF INCISIONAL HERNIA

- Infection uncontrolled
- Incision wrongly placed
- Improper suture material
- Increased intra-abdominal pressure

1. **Infection:** Cases operated for peritonitis such as perforated duodenal ulcer, gangrene of the intestines, etc. usually develop incisional hernia. The drainage tubes which are placed inside the peritoneal cavity help in reducing the postoperative incisional hernias, by draining peritoneal contents outside.
2. **Anatomical site:** The midline[1] is especially weak in the lower abdomen because of absence of posterior rectus sheath below the arcuate line or semilunar line.
3. **Obesity with weak muscle tone** predisposes to incisional hernia.
4. **Faulty technique** of closure of the abdomen or **faulty sutures** are also responsible for incisional hernia.
5. **Ascites, distension and persistent postoperative cough** further weakens the incision.
6. **Wrongly placed incisions** wherein nerves of the abdominal muscles are cut, precipitate incisional hernia. Lumbar incisions, lower midline incisions and large transverse incisions often give rise to incisional hernias (Figs 34.59 to 34.63).
7. **Incisions** wherein nerves are cut, the chances of incisional hernias are move. Examples: Subcostal incision for removal of gall bladder.

Fig. 34.59: Incisional hernia with decubitus ulcer due to obesity and weak muscle tone

[1] Is that the reason why we see many cases of incisional hernia after gynaecological operations?

Fig. 34.60: Incisional hernia following laparotomy for perforated duodenal ulcer. He had stormy postoperative period

Fig. 34.61: Incisional hernia—large (*Courtesy:* Dr CG Narasimhan, Senior Consultant Surgeon, Mysore, Karnataka)

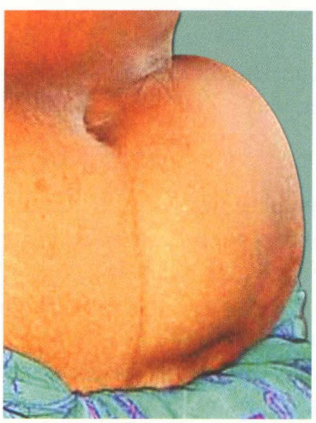


Fig. 34.62: Incisional hernia to the side (*Courtesy:* Dr Manjunath Shenoy, Professor of Surgery JSS Medical Collage, Mysore)

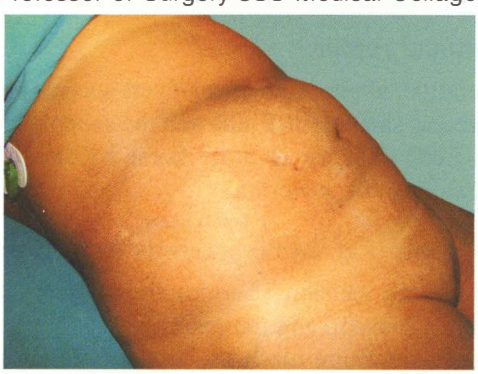


Fig. 34.63: Appendicectomy—incisional hernia

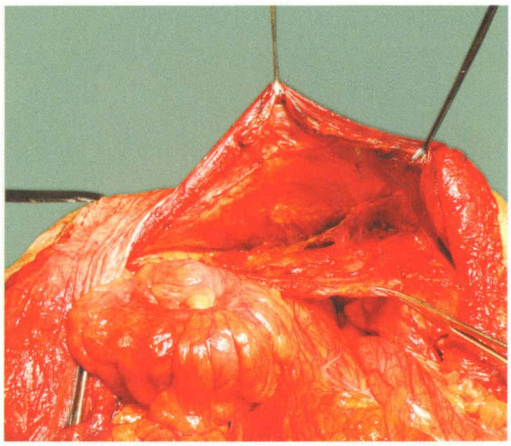


Fig. 34.64: Anatomical repair

Clinical features

- Serosanguinous discharge on the 4th postoperative day through the main suture line is a signal of development of partial or total wound dehiscence. Such cases later develop an incisional hernia.
- History of infection during the first surgery, postoperative cough is usually present.
- There is a bulge/swelling in relation to the scar.
- Scar is thin and evidence of secondary healing in the form of irregular scar may be present.
- Expansile impulse on cough and reducibility may be present.
- After reduction of the contents, a defect can be palpated through the scar. Defect depends upon number of stitches that have given way.

Treatment

- Surgical treatment is necessary if the **defect is narrow,** if there is **discomfort** to the patient or if there is a **danger** of obstruction (Figs 34.64 to 34.66).
- Preoperative preparation includes reduction of weight, control of cough, etc.
- There are various operations for the treatment of incisional hernia depending upon the size of the defect, anatomical location of the incision and presence of precipitating factors.

1. Anatomical repair

In this operation, all the anatomical layers such as peritoneum, posterior rectus sheath, linea alba and the subcutaneous tissue are identified. Closure is done layer by layer by using nonabsorbable suture material.

2. Mesh repair (Fig. 34.65 and refer Principles also)

- As most of the incisional hernias are due to a large defect in the main incision and the majority of it occurs in obese women, repair using mesh has become the most popular method. **Mesh repair is considered as the best repair, especially in obese, multiparous female patients with poor muscle tone.**
- In this operation, the sac is opened, greater omentum is excised, the contents are reduced followed by closure of the peritoneum. A mesh is kept in place which is sutured all around to the locally available tissues, without tension. Prolene mesh or Marlex mesh is commonly used. **In all these repairs, tensionless, nonabsorbable suture repairs are done.** The mesh is placed in the peritoneal space and covered by rectus muscles. Seroma is a common complication (Fig. 34.66).

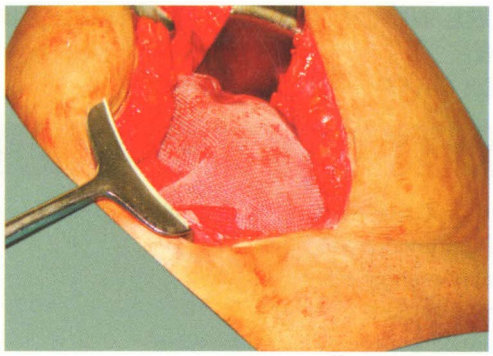


Fig. 34.65: Incisional hernia mesh repair

Principles

- Previous incision is opened to full length and the scar is excised.
- Flaps are raised on both sides
- The edges of rectus abdominis muscles are defined
- The sac is identified, dissected, freed from surrounding structures
- It is opened, contents reduced, may have to excise omentum
- Redundant sac is excised
- Peritoneum is closed
- Placement of the mesh: Onlay means subcutaneous. It is a simple procedure. Mesh should overlap at least 5 cm all around the defect. Otherwise, plane is created between posterior rectus sheath and rectus muscle, mesh is placed in that location and anterior rectus sheath is sutured. This is called retromuscular sublay mesh repair. This may not be possible below the arcuate line because there is no posterior rectus sheath there.

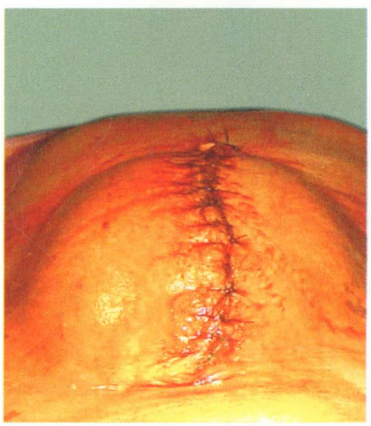


Fig. 34.66: Seroma after incisional hernia

Incisional hernia—laparoscopic repair

- Laparoscopic ventral hernia is the gold standard operation now.
- Major advantage is minimal scars, minimal pain, early recovery.
- It gives an excellent exposure from within
- One can define all the defects properly
- Reduce all the contents and a broad sac
- Defect can be sutured and broad mesh is placed
- Mesh which has absorbable surface will face the peritoneal cavity and nonabsorbable surface will face abdominal wall
- Transfascial sutures are put to fix the mesh (Figs 34.67 to 34.70).

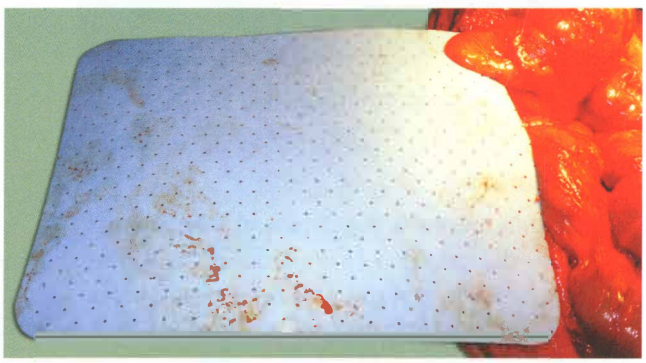


Fig. 34.67: Dual mesh

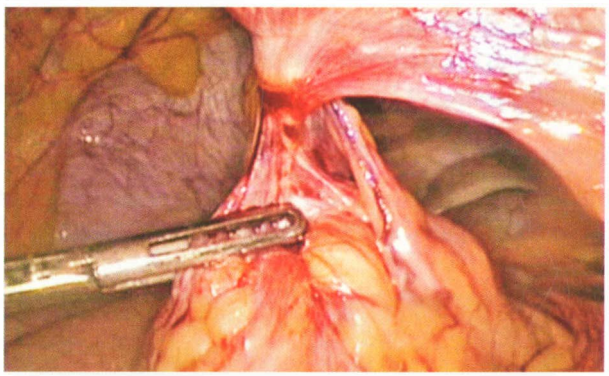


Fig. 34.68: Laparoscopic release of omentum from the hernial sac

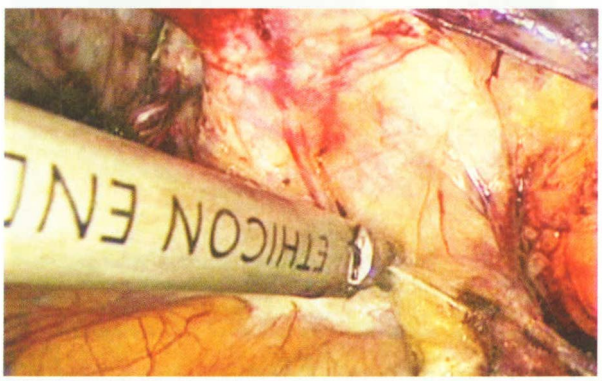


Fig. 34.69: Laparoscopic mobilisation of the sac

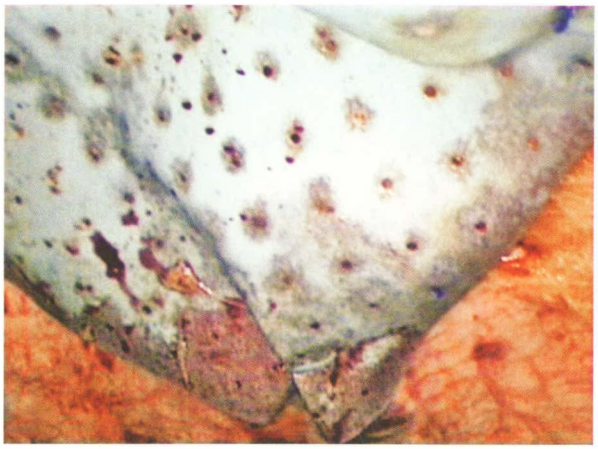


Fig. 34.70: Laparoscopic dual mesh has been kept in place and sutured

Management of a large incisional hernia (Fig. 34.71)

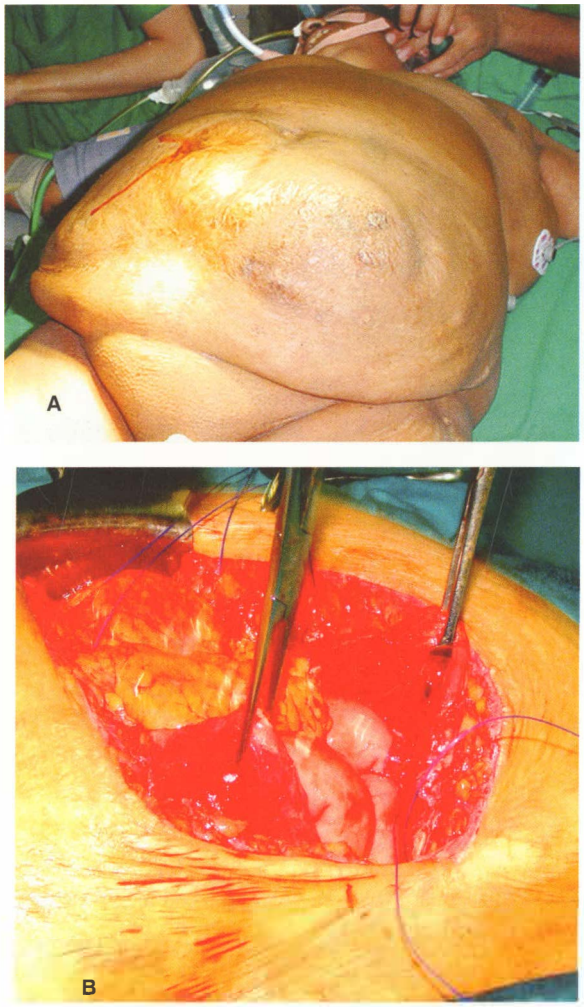
- CT scan is the best investigation to assess how much bowel is outside abdominal wall. If more than 25% is present outside, sudden reduction of the contents will result in compartment syndrome and pulmonary complications.
- Following advice or techniques are followed before taking up these cases:
  1. Fitness for surgery
  2. Chest physiotherapy
  3. Reduction of weight
  4. Deep vein thrombosis prophylaxis by giving low molecular weight heparin.
  5. Progressive pneumoperitoneum over weeks
  6. Additional lipectomy, removal of colon, omentectomy may have to be done.
  7. *Ramirez component separation technique*: In this, relaxing incisions are given in the external oblique aponeurosis or rectus sheath so that they can be brought together and stitched together because in these cases muscles are widely separated (Fig. 34.64).

3. Laparoscopic mesh repair

This is the procedure of choice today. A very good view of intraperitoneal contents and hernial sac is available once the telescope is introduced within the peritoneal cavity. All contents are reduced. A broad mesh can be placed in intraperitoneal or preperitoneal space depending upon its nature. Mesh is sutured all around. Recovery is fast and recurrence is very less.

4. The Keel operation

- It is also known as the Keel operation of Rodney-Maingot. It can be recommended for small hernias with good locally available tissues.
- In this operation, the sac is dissected and is pushed back into the abdomen ***without opening the peritoneum***.



Figs 34.71A and B: Three times operated case of incisional hernia, repair is being done

Inverting sutures are applied. **This repair on cross section *resembles the Keel of a ship.* This operation is considered obsolete now.**

EPIGASTRIC HERNIA

- It is also called **fatty** hernia of the linea alba.
- This type of hernia occurs in the epigastrium through the linea alba which extends between the xiphoid process and umbilicus.

Precipitating factors

- Sudden straining or heavy exercise results in the tearing of a few fibres of linea alba and is responsible for precipitating an epigastric hernia. Initially there is a small protrusion of extraperitoneal pad of fat. Rarely, if it enlarges, it is due to the dragging of the peritoneal sac. The opening is very narrow. Hence, the hollow viscus cannot enter the sac. Diastasis of rectus muscles which results in a wide linea alba can also precipitate an epigastric hernia.

Fig. 34.72: Epigastric hernia

Clinical features

- Common in muscular men, manual labourers.
- Typically the swelling is situated in the upper abdomen midway between xiphoid process and umbilicus. Often, it contains only an extraperitoneal protrusion of fat (Fig. 34.72).
- An expansile impulse on cough is rare.
- Dull aching pain is due to the fatty contents which are partially strangulated. However, tenderness is an important feature of epigastric hernia (Key Box 34.18).
- Many cases are associated with peptic ulcer disease.
- On head-raising, it becomes more prominent (Fig. 34.73).

Treatment

A small incision is made over the swelling and the fatty tissue is isolated. It is ligated and excised because usually a tiny blood

Fig. 34.73: Epigastric hernia: On head-raising, it becomes more prominent (head-raising test)

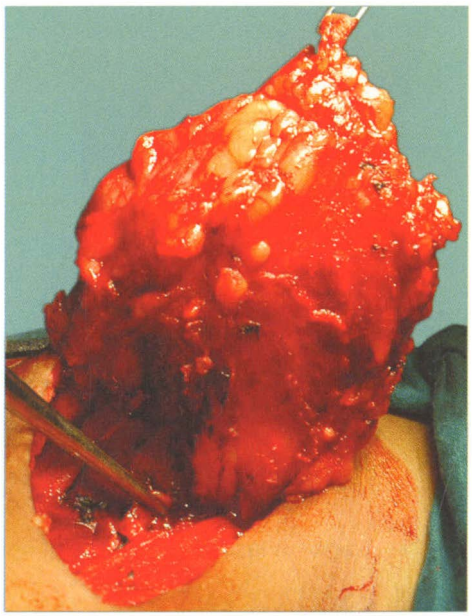


Fig. 34.74: Epigastric hernia sac

vessel enters the pad of fat. If a hernial sac is present, it is opened, the contents are reduced and the defect is closed using nonabsorbable sutures (Fig. 34.74).

KEY BOX 34.18

PECULIARITIES OF EPIGASTRIC HERNIA

- Common in muscular men
- Hernial sac is uncommon
- Hollow viscus in the sac is rare
- Impulse on cough is rare
- Reducibility is rare
- Tenderness is an important feature

RARE EXTERNAL HERNIAS

INTERPARIETAL HERNIA

- It is also known as interstitial hernia.
- Basically, they are inguinal hernias. However, the processus vaginalis sac instead of following the normal route into the scrotum, traverses between various layers of the abdominal wall (parietes) resulting in interstitial hernias.
- Patients with Down's syndrome and prune-belly syndrome are commonly affected.

Types

1. **Preperitoneal:** In this variety the hernial sac lies between the transversalis fascia and peritoneum. It is seen in about 20% of patients. The sac is like a small diverticulum.
2. **Interparietal:** It is also called as intermuscular type. It is the commonest variety wherein the sac passes between the external oblique and internal oblique muscles. The swelling caused by the hernial sac causes discomfort to the patient. Sometimes, this can be a bilocular sac.
3. **Extraparietal:** It is also known as inguino-superficial variety. In this variety the hernial sac passes exterior (superficial) to the external oblique aponeurosis beneath superficial fascia of the abdominal wall. It is commonly associated with undescended testis or ectopic testis.

- Majority of such cases present with features of intestinal obstruction.
- They are treated by identifying the sac, excision followed by closure of the defect or repair by using nonabsorbable sutures.

SPIGELIAN HERNIA[1]

- It is an interstitial hernia which occurs through the spigelian fascia. This is a thin strip of fascia which runs parallel to the outer border of rectus sheath from the tip of the 9th costal cartilage to the pubic tubercle.
- Since it is very wide in the region of umbilicus/arcuate line, spigelian hernias occur commonly at this level.
- Spigelian fascia contributes a few fibres to form rectus sheath.

Spigelian belt

- It is a 6 cm horizontal transverse zone located within umbilicus and 2 anterior superior iliac spine.
- Starts as a direct protrusion behind rectus abdominis.
- They are intramural; sac penetrates across transverse muscles and lies behind external oblique muscles.

Precipitating factors

Repeated pregnancies, advancing age, obesity, muscular degeneration, sudden strain due to coughing, weight lifting, etc. give rise to spigelian hernias.

Clinical features

- Seen in both sexes equally around 50 years of age.
- A round, soft, reducible swelling situated just below and lateral to the umbilicus—located typically at the junction of the arcuate line and lateral border of rectus abdominis. Sometimes, it is tender.
- The swelling gives rise to an expansile impulse on cough.
- As the hernia enlarges, it insinuates between external and internal oblique muscle. Hence, it is an example for interparietal hernia (Figs 34.75 to 34.78).

Investigations

1. An ultrasound can define the defect in the semilunar line.
2. X-ray abdomen, lateral view shows coils of bowel outside the peritoneal cavity.

[1] Prof Spigel, Professor of Anatomy and Surgery not only described spigelian fascia but also the caudate lobe of the liver.

Fig. 34.75: Obstructed spigelian hernia in a 70-year-old lady—first presentation to the hospital

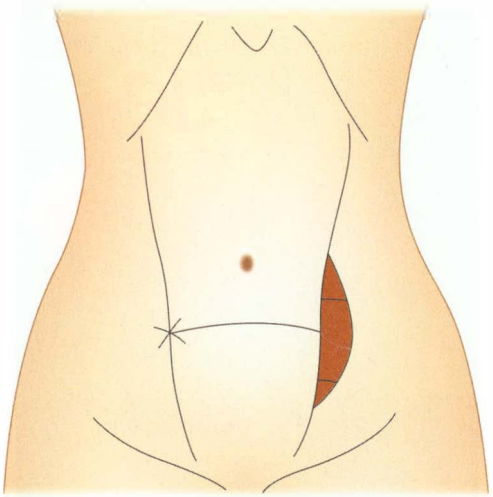


Fig. 34.76: Spigelian fascia and the site of hernia

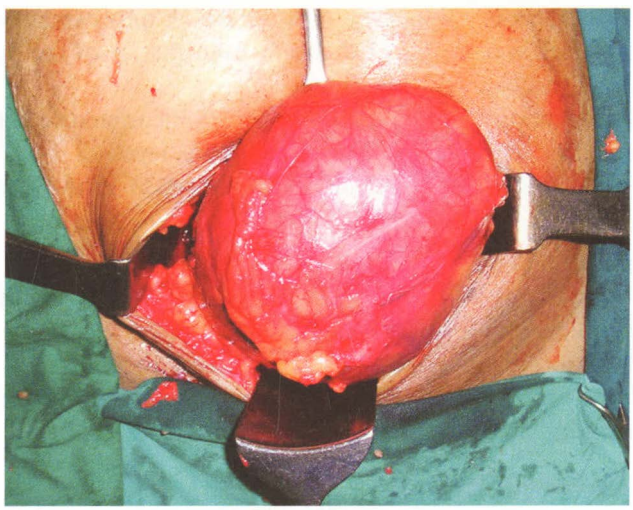


Fig. 34.77: Delivery of the sac

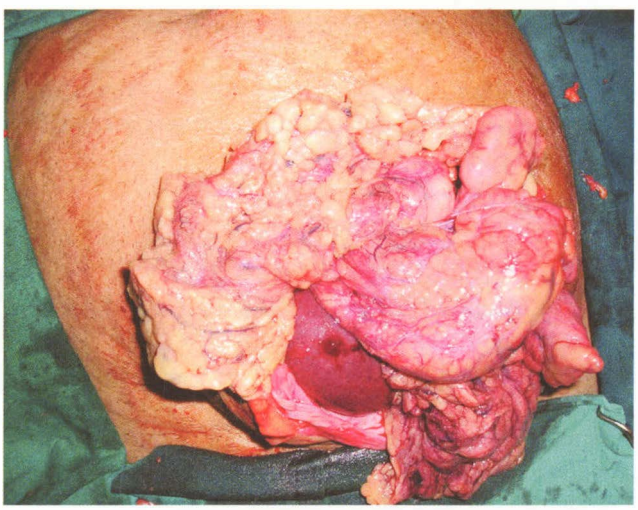


Fig. 34.78: Contents of the sac

(*Courtesy:* Prof MG Shenoy and Dr Prasad, KMC, Manipal, Figs 34.77 and 34.78)

Differential diagnosis

- Haematoma within the rectus sheath. However, it will not give rise to impulse on cough. It occurs suddenly and it will be a tender swelling.
- Pyogenic or pyaemic abscess can occur in the abdominal wall, more so, in diabetic patients. Tenderness and high temperature clinches the diagnosis.

Complication

Strangulation is common due to the rigid fascial ring surrounding the hernial sac. Richter's hernia also can occur here.

Treatment

An incision of about 5 to 6 cm is made over the swelling and abdominal wall muscles are split or cut. The sac is excised after reducing the contents and the defect is repaired. Recurrence occurs in about 5% of the patients.

LUMBAR HERNIA

Two types of lumbar hernia are well-recognised. They are as follows:

1. Primary which occurs through an anatomical defect:

- **Through the inferior lumbar triangle of Petit.** Its boundaries are:
  Inferiorly: Iliac crest
  Laterally: External oblique
  Medially: Latissimus dorsi
- **Through the superior lumbar triangle of Grynfeltt**. Its boundaries are (Fig. 34.79)
  Above: 12th rib
  Medially: Sacrospinalis
  Laterally: Internal oblique

2. **Secondary** to a renal operation done through a loin incision. It is an example of a *lumbar incisional hernia,* which occurs due to either infection or weakness of loin muscles. The operation done for tuberculosis of spine through a loin incision, very often gives rise to a secondary lumbar hernia (it is an incisional hernia) (Fig. 34.80).

Differential diagnosis

1. **Lipoma** is common in the lumbar region (loin). It is soft, lobular, and slips under the palpating fingers.
2. **Cold abscess** secondary to tuberculosis of the spine gives rise to a nontender swelling in the paravertebral space. Tenderness is present over the spine which gives a clue to the diagnosis. Patients may have deformity of the spine in the form of gibbus.

Treatment

Small defects can be closed with simple sutures. Large defects need to be closed with or without mesh.

PEARLS OF WISDOM

Primary lumbar hernias are very rare

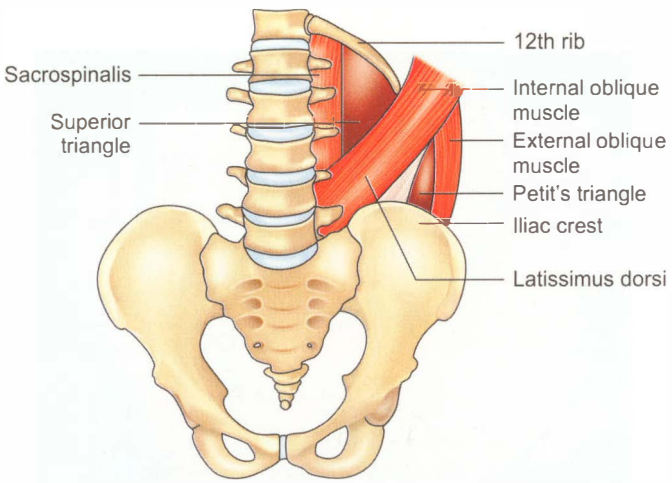


Fig. 34.79: Petit's triangle

Fig. 34.80: Lumbar hernia

OBTURATOR HERNIA

- This hernia occurs through the obturator canal which is bounded above by the superior ramus of pubis and below by the sharp edge of the obturator membrane.
- As the hernia is covered by the pectineus muscle, it is often overlooked.

Precipitating factors

- In females, the **obturator foramen** is wider in the transverse direction (it is triangular in shape in females and oval in males).
- Repeated pregnancies
- Loss of body weight
- Chronic lung diseases

Clinical features

- The **most common presentation** is acute intestinal obstruction **with strangulation** (80%). Recurrent attacks of intestinal obstruction which get resolved spontaneously is also common (Key Box 34.19).
- This hernia causes more pain than any other type of hernia. Pain often radiates along the obturator nerve and may even be referred to the knee *via* its geniculate branch called ***Howship-Romberg sign.*** The leg is usually kept in the *semiflexed position* and movement of the limb gives rise to pain. If the limb is flexed, abducted and rotated outwards the hernia becomes prominent. Patients are usually over 60 years of age and women are more frequently affected than men.
- Due to strangulation and blood in the hernial sac, bruising is seen below the medial edge of the inguinal ligament.
- A few patients (20%) complain of palpable ***hernial mass*** in the groin.
- Per vaginal examination can reveal a tender lump on the lateral side of the vault.

Treatment

The constricting agent in case of obstruction is the **obturator fascia**, which needs to be divided. Nerves and vessels are posterolateral to the hernial sac. Since majority of the cases present with intestinal obstruction and strangulation, a lower laparotomy is done. A grooved director is used to divide the obturator fascia.

KEY BOX 34.19

OBTURATOR HERNIA

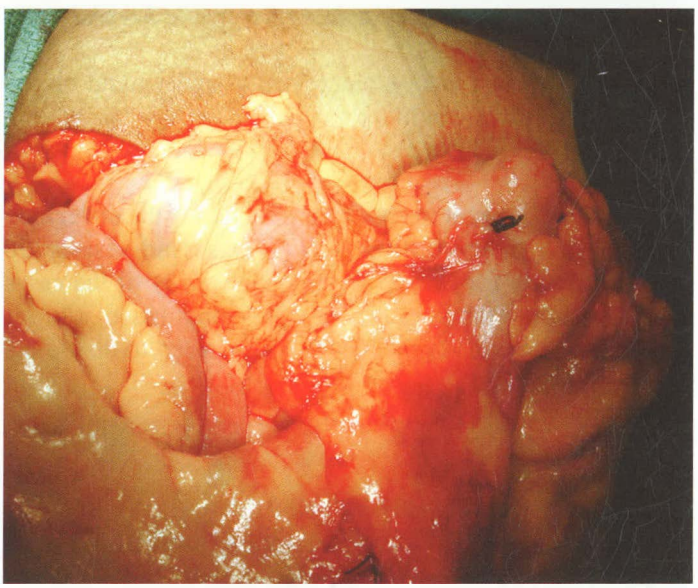
- The most common presentation is not a swelling but acute intestinal obstruction
- Can present as only pain in the knee—(Howship-Romberg sign)
- Vaginal examination: Tender mass can be felt on the lateral side
- Very high chance of strangulation

- The contents are reduced or if there is gangrene, the affected bowel is resected.
- Closure of the obturator opening is done by stitching the broad ligament over the opening or by using monofilament nylon.

PERINEAL HERNIA

These are very rare hernias which confuse many clinicians and present in a different varieties. Hernia protrudes through muscles and fascia of the perineal floor.

1. **Anterolateral perineal hernia**: This occurs in women and presents as a swelling of the labium majus. Often, the patient is examined by a gynaecologist and Bartholin's cyst is diagnosed (Figs 34.81 and 34.82).



Figs 34.81 and 34.82: A 40-year-old lady with large perineal hernia was diagnosed to have a Bartholin's cyst and was explored by a gynaecologist. To his surprise, it had 3 feet of small intestine and portions of large intestines. It started bleeding and he kept a drain and referred the patient to our hospital

2. **Posterolateral perineal hernia**: This type of hernia passes through levator ani and enters ischiorectal fossa.
3. **Median sliding hernia** is nothing but complete prolapse of rectum.
4. **Postoperative hernia** through perineal scar, e.g. after abdominoperineal resection wherein rectum is removed.
   - It also used to occur following perineal prostatectomy.

Clinical features

They can present as asymptomatic swelling, pain, dysuria, bowel obstruction or perineal ulceration and bleeding.

Diagnosis

- Ultrasound can detect loops of bowel/fluid.
- CT scan/MRI can clearly define the course of hernia, its relationship with urinary bladder/ureter and its descent into the pelvis.

Repair

It can be very difficult in large hernias. Often a combined approach, both perineal and abdominal, may be necessary. Mesh repair with adequate fascial and muscular perineal repair is required (Fig. 34.83).

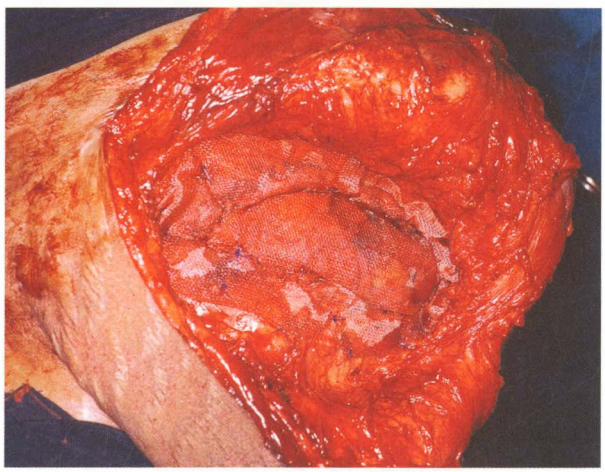


Fig. 34.83: Contents were reduced, sac was excised and a large mesh could be placed in the defect and sutured all around. Postoperatively she had urinary fistula. CT cystogram revealed bladder injury. It healed after two weeks

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been updated
- Inguinal anatomy is discussed in more detail
- Perineal hernia has been added
- European classification is added
- Biological mesh is added
- Special hernia is added
- New development in the repair of hernia such as hernia system and plug repair are added

MULTIPLE CHOICE QUESTIONS

1. Length of inguinal canal is:
A. 4 cm
B. 6 cm
C. 8 cm
D. 10 cm

2. Which was not a boundary of the Hesselbach's triangle?
A. Rectus abdominis
B. Inguinal ligament
C. Inferior epigastric artery
D. Testicular artery

3. Following are true about external ring *except:*
A. It is not a ring
B. It is a defect in the internal oblique aponeurosis
C. Invagination test is done through external ring
D. It transmits spermatic cord

4. About deep inguinal ring, which one of the following is true?
A. It is a defect in the external oblique aponeurosis
B. It is a defect in the internal oblique aponeurosis
C. It is a defect in the transversalis fascia
D. It is a defect in the cremasteric fascia

5. Following are true for deep inguinal ring *except:*
A. Deep ring is a defect in the transversalis fascia
B. Indirect hernia sac comes out lateral to the deep ring
C. Pantaloon hernia can be on both sides of deep ring
D. It is closed at the end of hernia repair to prevent recurrence

6. The ideal surgical treatment for sliding inguinal hernia will be:
A. Herniotomy
B. Bassini's herniorrhaphy
C Marsey's repair
D. Lichtenstein's repair

7. Following are contents of the spermatic cord *except*:
A. Vas deferens
B. Testicular artery
C. Genital branch of genitofemoral nerve
D. Ilioinguinal nerve

8. About sliding inguinal hernia following are true *except:*
A. Urinary bladder can be the part of hernial sac
B. It can be both direct or indirect type
C. The hernial sac should be twisted like as in indirect hernia treatment
D. Often it is irreducible

9. Richter's hernia refers to:
A. Hernia containing intestines
B. Strangulated hernia
C. Only a part of circumference of the intestine is caught in a hernial sac
D. Hernia containing urinary bladder

10. Immediate structure anterior to direct hernial sac is:
A. External oblique aponeurosis
B. Internal oblique aponeurosis
C. Transversalis facia
D. Posterior rectus sheath

11. Femoral hernia has following features *except:*
A. It is more common in women
B. It is below and lateral to pubic tubercle
C. It is known for strangulation
D. It can be managed by hernia truss

12. Diagnostic feature of a saphena varix is:
A. It is transilluminant
B. It is soft and reducible
C. It is below the pubic tubercle
D. Disappears on elevation of the leg

13. Which one of the following hernia is called Littre's hernia?
A. Hernia containing Meckel's diverticulum
B. Hernia containing urinary bladder
C. Hernia containing sigmoid colon
D. Hernia containing ovary

14. Which one of the following is the most important step in preventing recurrence of hernia?
A. Complete excision of cremasteric muscle
B. Reconstruction of the external ring
C. Reconstruction of the internal ring
D. High ligation of the sac

15. Following are true for strangulated inguinal hernia *except:*
A. It will be tense
B. Tender
C. Irreducible
D. Impulse on cough is present

16. Following are true for anatomy of the femoral canal *except:*
A. Femoral canal is the outermost compartment of femoral sheath
B. Femoral canal extends from femoral ring to saphenous ring
C. Femoral canal is below and lateral to pubic tubercle
D. Femoral canal contain lymph node of Cloquet

17. In obstructed femoral hernia, at surgery which of the following steps should not be done?
A. Best done with low approach through incision directly over the swelling
B. Closure of the ring is done by prolene suture material
C. Abnormal obturator artery should be looked for
D. Urinary bladder may be in danger

18. Following hernias are known for high chances of strangulation *except:*
A. Femoral hernia
B. Obturator hernia
C. Spigelian hernia
D. Direct hernia

19. In which condition femoral hernia occurs behind the femoral vessels?
A. Prune-belly syndrome
B. Poliomyelitis
C. Congenital dislocation of the hip
D. Defect in the lacunar ligament

20. In cases of epigastric hernia, all are true *except:*
A. It is more common in muscular men
B. Impulse on cough is common
C. Sac is uncommon
D. It is tender

21. Spigelian hernia is an example for:
A. Direct hernia
B. Indirect hernia
C. Interstitial hernia
D. Type of femoral hernia

22. In spigelian hernia swelling is seen:
A. Below and lateral to umbilicus
B. Below the umbilicus
C. Around the umbilicus
D. Just above the umbilicus

23. Differential diagnosis of lumbar hernia include following *except:*
A. Lipoma
B. Cold abscess
C. Haematoma
D. Meningocoele

24. Following are true for obturator hernia *except:*
A. Hernia is covered by pectineus muscle
B. Pain is radiated to the knee
C. Cannot be felt by vaginal examination
D. Patients keep their leg semiflexed

25. The most common presentation of obturator hernia is:
A. Groin swelling
B. Intestinal obstruction
C. Bruising below inguinal ligament
D. Tender mass in vaginal examination

ANSWERS

1 A	2 D	3 B	4 C	5 D	6 D	7 D	8 C	9 C	10 C
11 D	12 D	13 A	14 D	15 D	16 A	17 A	18 D	19 C	20 B
21 C	22 A	23 D	24 A	25 B					

35

Umbilicus and Abdominal Wall

- Classification of umbilical diseases
- Umbilical inflammation
- Umbilical fistulae
- Umbilical neoplasms
- Abdominal dehiscence
- Divarication of recti
- Rectus sheath haematoma
- Meleney's gangrene
- Desmoid tumour
- Endometriosis
- What is new?/Recent advances

CLASSIFICATION OF UMBILICAL DISEASES

I. Inflammation
  A. Omphalitis
  B. Granuloma
  C. Dermatitis
  D. Pilonidal sinus

II. Fistulae
  A. Faecal
    1. Patent vitellointestinal duct (Key Box 35.1)
    2. Carcinoma transverse colon
    3. Tuberculous peritonitis
  B. Urinary: Patent urachus
  C. Biliary

III. Neoplasms
  A. Benign
    1. Adenoma: Raspberry tumour
    2. Endometrioma
  B. Malignant
    1. Primary carcinoma
    2. Secondary carcinoma from: stomach, colon, ovary, and breast

IV. Umbilical hernia

V. Umbilical calculus (umbolith)

KEY BOX 35.1

UMBILICAL CORD STRUCTURES

IN FOETAL LIFE

- Umbilical vein
- Right and left umbilical arteries
- Urachus

IN EMBRYONIC LIFE

Vitellointestinal duct and structures mentioned above.

INFLAMMATION

A. Omphalitis

- Inflammation of the umbilical cord due to *Staphylococcus aureus* and Streptococci occurs in the neonatal period 3–4 days after birth. The incidence is increased in hospital births.
- Rarely, gram-negative organisms and *Clostridium tetani* can cause omphalitis[1], if strict aseptic precautions are not taken. If infection is not controlled, it can result in further complications.
  1. **Abscess of the abdominal wall:** Pus can be seen coming out of umbilicus. It may need drainage with antibiotic cover. Gentle squeezing will help, followed by antiseptic dressings and systemic antibiotics.
  2. **Extensive ulceration of the abdominal wall** similar to Meleney's ulcer, is a rare complication (subcutaneous synergistic gangrene) of omphalitis.

[1] Application of cowdung to the umbilical cord is still prevalent in a few places in our country. In addition to causing omphalitis, it can also cause neonatal tetanus.

3. **Septicaemia** can occur due to organisms entering the umbilical vein and then into portal vein. This results in pylephlebitis with jaundice, fever, chills and rigors.
4. **Neonatal jaundice** due to intrahepatic cholangitis.
5. **Portal vein thrombosis** resulting in extrahepatic portal hypertension (prehepatic).
6. **Umbilical hernia** can occur due to a weak scar produced by sepsis.

B. Granuloma

Granuloma indicates persisting inflammation underneath. It is a cause of great concern and worry to the patients. This can be destroyed by **application of copper sulphate or silver nitrate solution**.

C. Dermatitis

Dermatitis more often occurs in adults wherein chronic infection of the umbilicus sets in with foul smelling discharge.

D. Pilonidal sinus

Umbilicus is a low area compared to the surface of abdominal wall. Hence, hairy men may shed their hair which accumulates in the umbilicus and may result in pilonidal sinus. It may need removal of sinus along with tuft of hair or rarely the umbilicus itself.

UMBILICAL FISTULAE

A. Faecal

1. **Persistent vitellointestinal duct** is an uncommon congenital anomaly. Many a time the intestinal opening is so small that only mucoid contents come out of umbilicus. Rarely, if the opening is big, omphaloenteric faecal fistula results.
2. **Internal hollow viscus malignancies,** especially carcinoma of the transverse colon can erode through umbilicus resulting in a faecal fistula.
3. **Tuberculous peritonitis** induces dense adhesions, strictures and perforations. A perforation which is sealed off by coils of matted bowel and omentum results in local abscess which may perforate through a weak point, i.e. umbilicus resulting in a faecobiliary fistula. If a diagnosis can be proved by wall biopsy of the sinus/fistula antituberculous treatment can cure the disease. Laparotomy is extremely difficult in such cases. One may end up creating more holes in the bowel and is better avoided.

PEARLS OF WISDOM

One innocent disease (V I DUCT), one prevalent disease (TB) and one malignant disease (carcinoma bowel) are the causes of umbilical faecal fistula.

B. Patent urachus (Fig. 35.1 and Key Box 35.2)

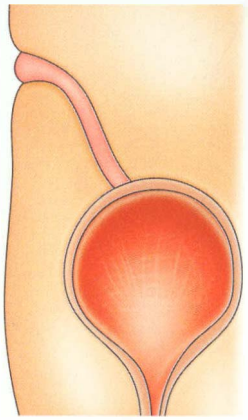


Fig. 35.1: Patent urachus

- The ventral urogenital sinus which forms the urinary bladder is continued cranially as urachus which extends into the umbilical cord—allantoic stalk. If this portion persists, patent urachus forms which connect umbilicus with urinary bladder. If it is fibrosed, as it occurs normally, it is called **median umbilical ligament**.
- A patent urachus may manifest as urinary discharge from umbilicus. It manifests usually in childhood and early adult life. In most cases, there will be some kind of obstruction to the normal passage of urine. Entire urachus is excised after correcting distal obstruction.

KEY BOX 35.2

ABNORMALITIES OF URACHUS

- Patent urachus
- Urachal sinus
- Urachal cyst
- Urachal diverticulum

C. Biliary fistula

Rarely, perforation of gall bladder due to severe form of cholecystitis may result in local abscess which may rupture through umbilicus resulting in biliary fistula. Instances are recorded wherein stones have come out of umbilicus.

NEOPLASMS

1. **Umbilical adenoma** is a pedunculated swelling having raspberry colour. Hence, the name **Raspberry tumour**.
   - It is due to unobliterated vitellointestinal duct.
   - Mucosa of the persistent duct prolapses through umbilicus and produces this adenoma.
   - It is moist with mucus and tends to bleed (columnar epithelium rich in goblet cells).

   Treatment

   - If the tumour is pedunculated, a ligature is tied around it and by a few days, the adenoma drops off.
   - If tumour reappears, excision of umbilicus is advised.
2. **Endometrioma** of umbilicus is rare but patients have a typical history to tell, i.e. it bleeds during menstruation.
3. **Malignant:** Secondary carcinomatous nodule in and around umbilicus reflects advanced malignancy commonly from stomach and colon. Ovary, uterus, breast are other causes.

SOME INTERESTING DISEASES OF UMBILICUS

Fig. 35.2: Sister Mary Joseph's nodule

Fig. 35.3: Exomphalos major

Fig. 35.4: Irreducible umbilical hernia in a cirrhotic patient

Fig. 35.5: Umbilical hernia

Fig. 35.6: Pilonidal sinus

Fig. 35.7: Observe skin of umbilical hernia in cirrhosis of liver, resembling scrotal skin

The nodule is tender, fixed and reddish in colour (Fig. 35.2). Figures 35.3 to 35.7 show some interesting diseases of umbilicus.

UMBILICAL HERNIA

Umbilicus is one of the weak points in the body. Hence, it is one of the sites of hernia. All the details of umbilical hernia have been discussed in page 566.

UMBOLITH

- It is composed of desquamated epithelium which becomes inspissated and gets collected in the umbilicus. With secondary infection, there will be blood-stained discharge. It is treated by controlling infection, debridement and if necessary removal of umbilicus.
- This umbolith or umbilical calculus is black in colour.

Fig. 35.8: Abdominal wall abscess in a diabetic patient (*see* clinical notes)

CLINICAL NOTES

A 60-year-old diabetic, male patient had a large pyaemic abscess on the abdominal wall below and to the right of umbilicus. An incision and drainage of the abscess was done. Within 2 hours of surgery, the surgeon was called to see this patient who had hypotension and blood was pouring out of the incision. Exploration of the wound revealed large 'clots' and bleeding from inferior epigastric artery, which was ligated. Probably, while breaking all the loculi of the abscess cavity, the vessel was injured.

ABDOMINAL WALL

PYOGENIC ABSCESS (Fig. 35.8)

Abdominal wall is one of the sites of pyogenic abscess especially in diabetic patients. It is a part of pyaemia. Localised tenderness suggests an abscess. Diagnosis can be confirmed by ultrasound and it is treated by incision and drainage.

ABDOMINAL WALL VEINS (Fig. 35.9)

- Veins are seen in portal hypertension. In relation to umbilicus, they are called as **caput medusae**. Direction of

Fig. 35.9: Dilated tortuous veins due to portal hypertension—caput medusae

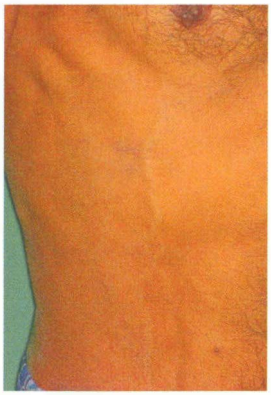


Fig. 35.10: Veins on the lateral abdominal wall suggestive of inferior vena caval obstruction

Fig. 35.11: Wound dehiscence in a case of APR (abdomino perineal resection)—probably due to placement of colostomy closer to the suture line

the veins are important which can be demonstrated by emptying the vein and filling.

- In cases of inferior vena caval obstruction, veins are seen on the flank. These veins are called as inguino-axillary veins (Fig. 35.10).

BURST ABDOMEN: ABDOMINAL DEHISCENCE

A soundly healed abdominal scar can withstand any amount of intra-abdominal pressure. However, 1–2% of the abdominal wounds (incisions) give way resulting in prolapse of intra-abdominal contents outside.

This causes great concern, or anxiety to the patient, and more so for relatives. It is said that the anxiety and worry caused by the intestines prolapsing out is much more than that is caused by emergency re-explorations for open cardiac surgery. It is not possible to prevent wound dehiscence totally because causative agents are multifactorial.

Factors responsible for wound dehiscence (Key Box 35.3)

1. **Surgery:** It depends upon the type of surgery done. Surgery done for grossly contaminated cases such as peritonitis, biliary fistula or faecal fistula have a high incidence of wound dehiscence (Fig. 35.11).

KEY BOX 35.3

BURST ABDOMEN: FACTORS

- **S**urgery → Peritonitis
- **S**epsis → Uncontrolled infection
- **S**utures → Absorbable—catgut
- **S**urgeon → Poor quality
- **S**ick patient → Malignancy, diabetes, uraemia, jaundice
- **S**training → Coughing, vomiting

Remember the causes of burst abdomen as **6S.**

2. **Sepsis:** Uncontrolled infection (sepsis) can digest the suture material used and will result in burst abdomen.
3. **Suture material used:** Absorbable sutures such as catgut give rise to increased incidence of wound dehiscence than nonabsorbable sutures.
4. **Surgeon-related factors:** Meticulous dissection, haemostasis, gentle handling of tissues, a good tensionless tight closure, carefully judged incisions will have reduced incidence of burst abdomen. Midline vertical incisions have decreased chance of wound dehiscence than paramedian incision.
5. **Sick patient:** Patients with malignancy, jaundice, obesity, anaemia, hypoproteinaemia, uraemia have poor wound healing.
6. **Straining:** In the postoperative period, violent cough, persistent vomiting, abdominal distension due to paralytic ileus predispose to burst abdomen.

Clinical features

- Patients who are recovering reasonably well in the postoperative period suddenly complain of pink or brownish coloured serosanguinous discharge. It is the pathognomonic sign of burst abdomen.
- It usually occurs on the 6th to 8th postoperative day.
- If skin sutures are removed, omentum or small bowel coils will be seen outside.

PEARLS OF WISDOM

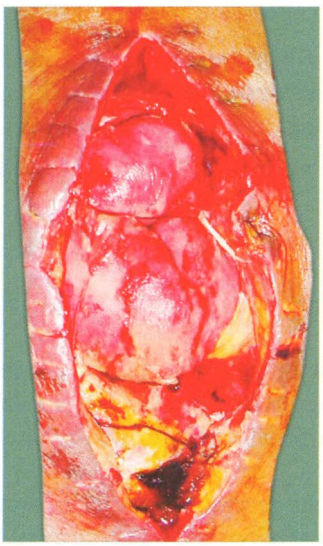
Interestingly, it is a painless, shockless disruption (with) full of apprehension.

Treatment

- Reassurance
- The bowel or the contents are covered with pads and bandage.
- Emergency surgery and closure is done.

Principles of surgery

1. Adequate exposure
2. Bowel is washed with saline and gently replaced into the peritoneal cavity.
3. Edges of the wound/incision are trimmed.
4. A single layer closure of the abdominal layer, by taking suture bites through whole thickness of the abdominal wall is done.
   - A few tension sutures (Figs 35.12 and 35.13) tied over a rubber or a plastic tube are placed and are removed after 2 weeks.
   - It should be remembered that **secondary wound healing is better than primary wound healing** and infection rarely occurs.



Figs 35.12 and 35.13: Wound dehiscence due to gangrene of the intestine—laparotomy was done, resection followed by closure of the abdomen with tension sutures

5. **Closer of midline incision:** In all cases of midline incision it is the linea alba with or without anterior fascial layer is approximated to the corresponding fascia on the other side.
   - "Fascia" usually refers to the anterior rectus fascia, the fascia above the rectus muscles. This fascia holds the abdomen together and is the most important layer of closure.
   - The fascia can extend beyond the muscles and bind to other fascia.
   - An extension of the fascia is called an aponeurosis.

Complications of wound closure

I. Early complications:
- Infection
- Dehiscence

II. Late complications:
- Incisional hernia
- Suture sinus
- Wound pain

Prevention of incisional hernia

1. Use correct suture technique

- Continuous suture with self-locking knot(s)
- Suture length (SL) : width length (WL) ratio > 4
- Small stitches at close intervals
- Aponeurosis only
- Minimal tension
- Monofilament, slowly absorbable suture

2. Prevent wound infections

- Correct suture technique
- Attention to hygiene measures
- Additional measures, e.g. antibacterial sutures

3. Surgeon-related factors

See Key Box 35.4

KEY BOX 35.4

SURGEON'S ROLE IN PREVENTING INCISIONAL HERNIA

- Continuous *vs* interrupted sutures
- Knotting technique—secure knots
- Suture length/wound length ratio
- Bite size
- Mass closure *vs* aponeurosis only
- Tensionless sutures
- Suture material—nonabsorbable
- Anti-SSI (surgical site infection) measures—prophylactic antibiotics

4. Continuous sutures

See Key Box 35.5

KEY BOX 35.5

CONTINUOUS SUTURES

- Continuation suturing is faster
- Accommodates wound lengthening due to distension
- Bursting strength of wound is significantly higher
- Minimises number of knots—equivalent or lower incidence of incisional hernia
- Disadvantages (theoretical): Wound security depends on single strand of suture and limited number of knots.

DIVARICATION OF RECTI

- In this condition, the two rectus abdominal muscles are widely separated (not in the midline).
- Repeated pregnancy in quick succession is the most important cause. Chronic constipation or overstraining may be another factor. Obviously women are commonly affected.
- Exercises and abdominal corset are helpful.
- Symptomatic cases are operated—divaricated recti are brought towards midline.

RECTUS SHEATH HAEMATOMA

Collection of blood in relation to rectus sheath and muscles occurs due to tearing of one of the branches of inferior epigastric artery. A parietal haematoma occurs usually at the level of the arcuate line. It is an uncommon condition. However, the causes can be as follows:

1. **Trauma:** A sudden blow to the abdominal wall.
2. **Straining:** Sudden straining such as violent cough or vigorous exercise in a muscular man can cause haematoma.
3. **Pregnancy:** Rarely, the cause of haematoma can be pregnancy, in late trimester. The exact cause is not known.

Clinical features

- History of sudden straining or coughing, etc.
- A tender lump develops just below and to the side of umbilicus at the level of arcuate line where posterior rectus sheath is absent.
- Nausea, vomiting, pyrexia are the other features.

Differential diagnosis

Spigelian hernia (it is rare)

Treatment

- The condition is self-limiting. With antibiotics and analgesics, a haematoma subsides within 5–7 days.
- If it persists or progresses or if there is a doubt about the diagnosis, exploration and evacuation of haematoma should be done and the bleeding vessels are ligated. The results and recovery are excellent.

MELENEY'S PROGRESSIVE POSTOPERATIVE SYNERGISTIC GANGRENE

This dangerous complication is rare nowadays, thanks to the good pre- and postoperative antibiotics.

Aetiopathogenesis

- It is caused by synergistic action of microaerophilic non-haemolytic Streptococcus and *Staphylococcus aureus*.
- Surgical operations which have increased risk of Meleney's gangrene include perforated appendix, biliary tract surgery, colectomy, etc.
- Atherosclerosis, diabetes are the other precipitating factors.
- Starts as cellulitis with reddish skin and postoperative fever.
- The spread may occur within 3–5 days, with extensive gangrene and sloughing of the skin of the abdominal wall with purulent discharge.

Clinical features

- Postoperative patient with cellulitis of abdominal wall
- Fever of moderate degree, an extremely tender abdominal wall and purulent discharge.
- Toxicity and deterioration of general health may follow soon.

Treatment

1. **At the stage of cellulitis:** Broad spectrum antibiotics to cover not only the organisms mentioned above but to cover anaerobic organisms. Thus, a combination of benzyl penicillin, gentamicin and metronidazole is used.
2. **At the stage of gangrene:** Emergency aggressive debridement is the treatment. Dead skin and subcutaneous tissue is excised, pus drained and the slough is removed.
3. **Hyperbaric oxygen** may be very useful.
4. Skin grafting is done, once the wound is healed with granulation tissue.

FIBROMATOSES: DESMOID TUMOUR

Classification

- *Superficial or deep*. **Deep** fibromatoses, also called ***Aggressive fibromatoses and desmoid tumours*** is an **uncapsulated fibroma** which occurs in the abdominal wall.
- It arises from muscles and aponeurotic layer of the abdominal wall.

Incidence

- In children, most desmoid tumours are extra-abdominal with a female predominance.
- In young adults desmoid tumours almost always occur in the abdominal wall (of women). ***Hormonal effects and pregnancy*** are believed to influence the growth of this tumour.
- Some tumours express hormone receptors (oestrogen and progesterone), and therefore ***tamoxifen*** and other hormonal modulators are among the adjuvant therapies for this tumour.

Aetiopathogenesis

- Childbirth, trauma or operative scars are the possible aetiological factors.
- Desmoid tumour is one of the components of Gardner's syndrome.
- It is benign but has a tendency to infiltrate the muscles. Some fibromas exhibit dysplastic changes. The cut surface is compared to an onion—whorled fibroma with spindle-shaped cells.
- Sarcomatous changes and metastasis do not occur.

Clinical features

- **Abdominal fibromatosis** is far less prone to recurrences than desmoid tumours in other sites.
- It usually occurs in the abdominal wall of women of childbearing age during or after pregnancy.
- Clinically the lesions present as deep-seated, firm, nonencapsulated, slow-growing, locally invasive and painless masses.

- It typically manifests as a slow-growing, progressive mass that becomes more prominent on abdominal muscle contraction. Mass is in the abdominal wall and is firm to hard in consistency.
- **Mesenteric fibromatosis**, probably the commonest among the group, usually presents as a slow-growing mass that involves small bowel mesentery or retroperitoneum.
- The recurrence rate of mesenteric fibromatosis seems to be substantially higher in patients who have Gardner's syndrome than in patients who do not have this syndrome.

Treatment

- Simple excision results in recurrence. Hence, **wide excision with 2–3 cm** of normal **healthy margins** is necessary with reconstruction of the abdominal wall (Figs 35.14 and 35.15).
- In spite of adequate surgery, 10–20% chances of recurrence occurs (Key Box 35.6).

Fig. 35.14: Wide excision of recurrent desmoid tumour in the abdominal wall

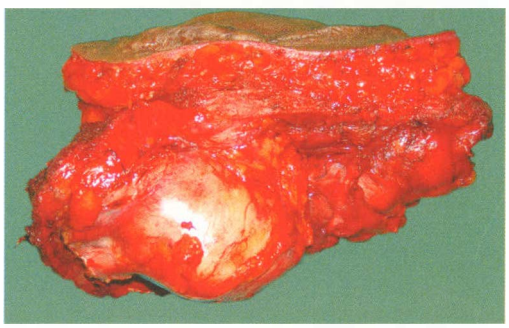


Fig. 35.15: Specimen of desmoid tumour which is removed along with normal tissue

ENDOMETRIOSIS OF THE ABDOMINAL WALL

Drugs like sulindac and tamoxifen have also been used here, with some success.

- It occurs due to mechanical implantation of endometrial cells during surgery (sites—Key Box 35.7 and Fig. 35.16).
- Painful, palpable swelling, more symptomatic at the time of menstruation are characteristic features.
  - Perimenstrual cyclical bleeding can occur
- Oral contraceptive pills may control the symptoms.
- Otherwise, excision of the nodule has to be done (Fig. 35.17).

KEY BOX 35.6

PECULIARITIES OF DESMOID TUMOUR

- Uncapsulated fibroma
- Infiltrates muscles, even though benign
- Does not change into sarcoma
- It may be a part of Gardner's syndrome
- Simple excision results in recurrence
- Wide excision is recommended

KEY BOX 35.7

SITES OF ENDOMETRIOSIS

- Laparoscopic port – Umbilicus
- Gynaecological surgery – Abdominal incision
- Episiotomy – Perineum

Fig. 35.16: Scar endometriosis

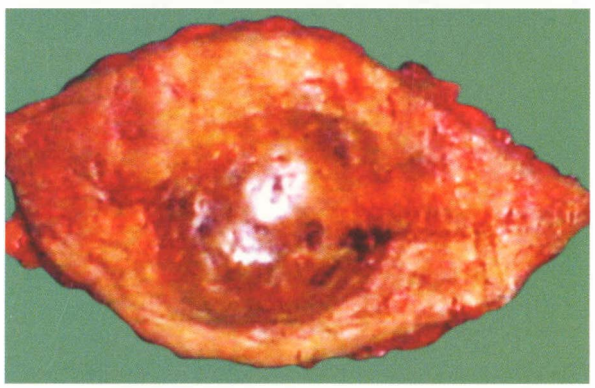


Fig. 35.17: Wide excision of scar endometriosis

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- Desmoid tumour and fibromatosis have been updated.
- A few coloured photographs have been added.

MULTIPLE CHOICE QUESTIONS

1. The following is *not* a cause of umbilical faecal fistula:
A. Persistent vitellointestinal duct
B. Tuberculosis
C. Carcinoma
D. Raspberry adenoma

2. The following statement is true about burst abdomen:
A. It is very painful
B. It is associated with shock
C. Straining can produce it
D. Conservative management is the choice

3. About desmoid tumour:
A. Is a capsulated fibroma
B. Infiltrates muscles
C. Often changes to sarcoma
D. Simple excision is recommended

4. The following is true of Meleney's postoperative synergistic gangrene *except*:
A. Is progressive
B. Can occur after appendicectomy
C. Starts as cellulitis
D. Is painless

5. The following is true about rectus sheath haematoma *except*:
A. It can occur with pregnancy
B. Sudden straining can precipitate it
C. Presents as painless lump above umbilicus
D. Nausea, vomiting and fever are other features

6. The principles of surgery for burst abdomen includes all of the following *except*:
A. Layer by layer careful suture
B. Adequate exposure
C. Tension sutures
D. Trimming of edges

7. The pathognomonic sign of burst abdomen is:
A. Shock
B. Pink or brown serosanguinous discharge
C. Pain
D. Occurs on the second postoperative day

8. Umbilical adenoma:
A. Sessile swelling
B. Is a premalignant condition
C. Can be treated with ligature
D. Occurs due to persistent umbilical vessels

9. Pilonidal sinus is known to occur in the following places *except*:
A. Internatal cleft
B. Umbilicus
C. Interdigital cleft
D. Axilla

10. Meleney's gangrene is a synergistic gangrene caused by:
A. Streptococcus and Staphylococcus
B. *E. coli* and *Klebsiella*
C. Clostridium and Pseudomonas
D. *Salmonella typhi* and *paratyphi*

ANSWERS

1 D	2 C	3 B	4 D	5 C	6 A	7 B	8 C	9 D	10 A

36

Blunt Abdominal Trauma, War and Blast Injuries and Triage

- Liver injuries
- Small bowel injuries
- Colonic injuries
- Duodenal injuries
- Pancreatic injuries
- Renal injuries
- Retroperitoneal haematoma
- Blast injuries
- Warfare injuries
- Missile wounds of abdomen
- Triage
- What is new?/Recent advances

Introduction

Blunt abdominal trauma (BAT) is one of the common surgical emergencies encountered by general surgeons. Increasing number of vehicles, high speed and poor maintenance of the roads are the contributing factors. Blunt injury abdomen with polytrauma is one of the commonest causes of death in the younger population. Thus, it is important for a house officer to recognise a polytrauma patient, to diagnose and to suspect intra-abdominal injury, so that urgent resuscitation and treatment can be offered to the patient at the proper time, at the proper hospital and by a proper surgeon. Major systems involved are given in Key Boxes 36.1 and 36.2.

Craniospinal and chest injuries are discussed in their respective chapters. Pelvic and skeletal injury is beyond the limits of this book. In this chapter, blunt injury of the abdomen is discussed.

KEY BOX 36.1

COMMON VISCERA INVOLVED IN BLUNT INJURY

• Spleen	Significant bleeding
• Liver	Significant bleeding
• Kidney	Significant bleeding
• Intestines	Perforation—peritonitis
• Pancreatico-duodenal injuries	Usually missed—bleeding
• Diaphragm	Missed—tachypnoea
• Urinary bladder	Urinary peritonitis

KEY BOX 36.2

MAJOR SYSTEMS INVOLVED

- Craniospinal
- Chest
- Abdomen
- Pelvis
- Skeletal

Causes of blunt injury abdomen

1. Rail and road traffic accidents (most common)
2. Fall from a height and dashing against an object
3. Seat belt syndrome
4. Assault

Mechanism of injury/pathophysiology

- Vehicular trauma is by far the leading cause of blunt abdominal trauma.
- Broadly BAT can be explained by **three mechanisms:**
  1. **Rapid deceleration:** It causes differential movement among adjacent structures. Shearing forces are created; they cause solid, visceral organs and vascular pedicles to tear at relatively fixed points of attachment. Examples:
     - Renal pedicle injury
     - Injury to distal aorta than proximal mobile aorta as former is attached to thoracic spine.
  2. **Crushing effect:** Here solid viscera are crushed between anterior abdominal wall and vertebral column or posterior thoracic cage.

3. **Sudden dramatic rise in the intra-abdominal pressure** due to external compression
   - Hollow viscus ruptures (in accordance with principles of **Boyle's law**).

Investigations

1. **Complete blood count**, coagulation studies, grouping and cross-matching. Fall in haemoglobin is an indication of ongoing haemorrhage—especially while managing a patient with liver/splenic injury on conservative line of management.
2. **Serum electrolyte** analysis
3. **Serum amylase/lipase**
   - May be elevated because of **pancreatic ischaemia due to hypotension**
   - **Persistent elevation** may be indication of **intra-abdominal injury.**
4. **Plain X-rays** (Fig. 36.1)
   - Chest X-ray: Pneumoperitoneum—fundic, stomach (air bubble in thorax as in diaphragmatic injury, retroperitoneal air—duodenal perforation.
   - Pelvic fractures
5. **Role of ultrasound** (Fig. 36.2)
   FAST: Focussed assessment with sonography for trauma
6. **Diagnostic peritoneal lavage (DPL)**
   It is indicated in BAT in following situations:

- Multiple injuries and shock
- Spinal cord injury
- Obtunded patient with possible abdominal injury
- Intoxicated patient

Also *see* splenic trauma (page 619).

Types

- **Open:** Infraumbilical skin incision and open peritoneum.
- **Semiopen:** Infraumbilical skin incision deepen up to linea alba.
- **Closed:** Blind insertion of needle.

Fig. 36.1: Chest X-ray showing diaphragmatic hernia on the left side

Precautions

- Foley's catheter to empty bladder
- Ryle's tube to empty stomach
- X-ray pelvis to detect pelvic fracture.

Positive DPL

- 10 ml of gross blood aspirate before infusion of lavage fluid.
- More than 100,000 RBC/ml
- More than 500 WBC/ml
- Bile and bacteria are demonstrated
- Vegetable matter

PEARLS OF WISDOM

Positive DPL means intraperitoneal injury is present. It does not mean that the patient should be shifted to operation theatre immediately.

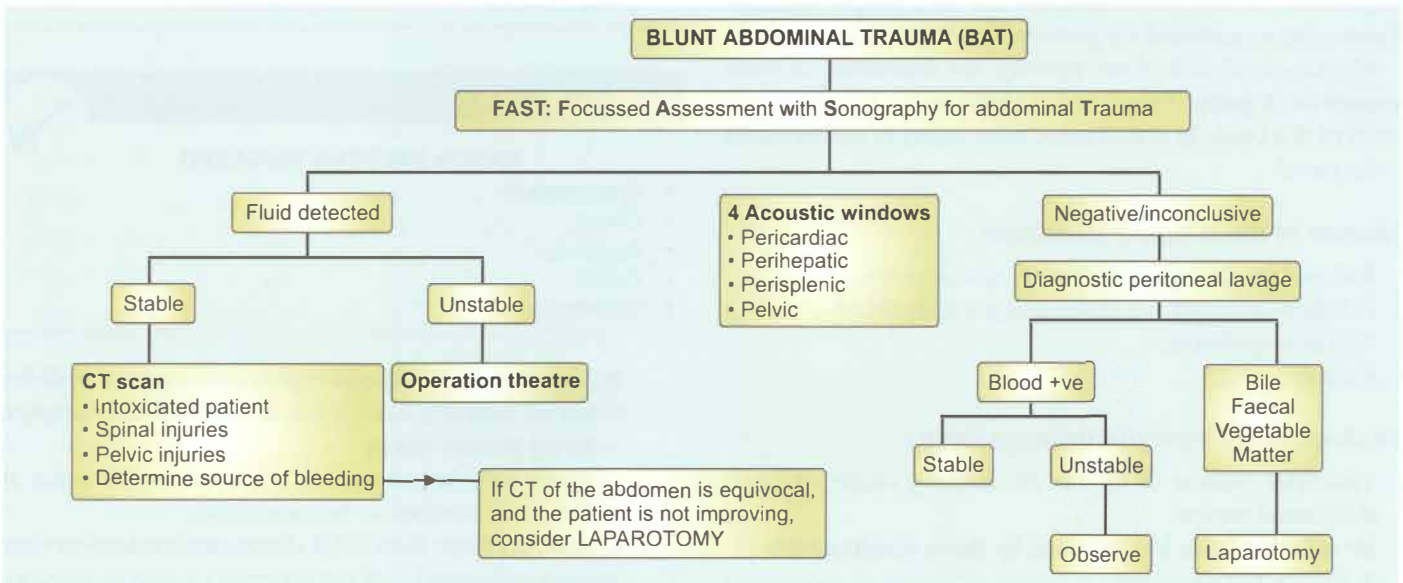


Fig. 36.2: Algorithm of investigations in BAT (*see* FAST)

7. CT scan (Fig. 36.3)

- Gold standard for solid organ injuries
- CT also can reveal other associated injuries such as vertebral or pelvic fractures.
- CT can also pick up **diaphragmatic injury** (CT chest).
- It can detect **source of haemorrhage**
- CT is an excellent scan for pancreas, duodenum, etc.

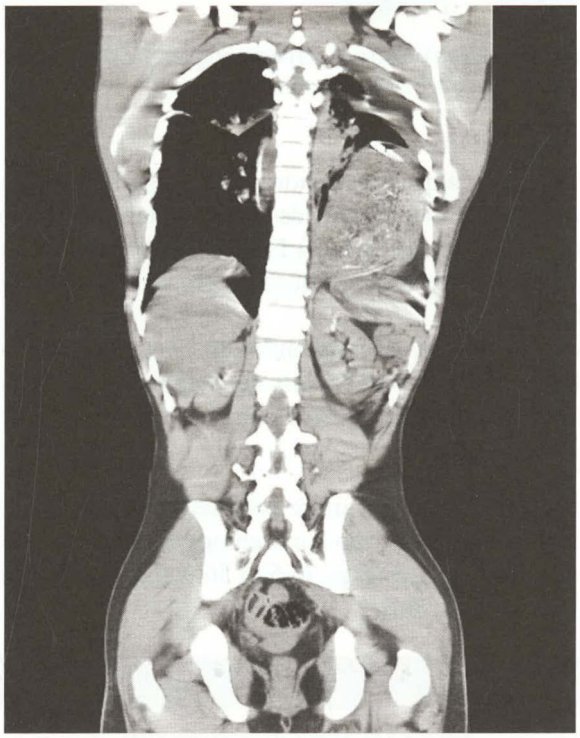


Fig. 36.3: Diaphragmatic hernia—CT scan (*Courtesy:* Dr Yashdeep Sharma, Associate Professor, KMC, Manipal)

8. Diagnostic laparoscopy: Done when CT scan is negative, suspicion of diaphragmatic injury is present (Fig. 36.4).

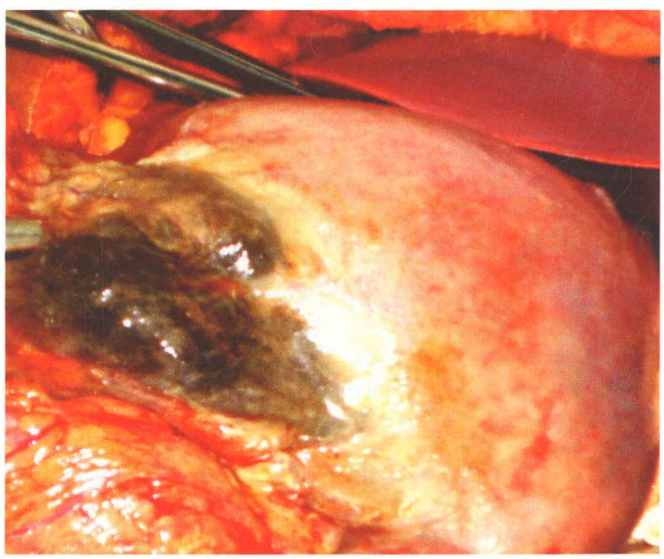


Fig. 36.4: Diaphragmatic hernia with gangrene of the stomach

PEARLS OF WISDOM

Stable patients with solid organ injuries are managed more often by nonsurgical methods with close monitoring.

Remarks

With the availability of FAST and CT scan, role of DPL is now limited to unstable patients whose FAST results are negative or inconclusive.

ON ARRIVAL AT CASUALTY

Primary survey—to identify and treat life-threatening injuries

ABCDE (ATLS protocol)	AMPLE (History)
Airway	Allergy
Breathing	Medication
Circulation	Past medical illness
Disability	Last meal
Exposure	Events leading to incident

Secondary survey—physical examination

- **Initial clinical assessment in an alert patient:** Pain, tenderness, gastrointestinal haemorrhage, hypovolaemia. Clinical evaluation alone has an accuracy rate of only 65% for detecting blood in peritoneal cavity.
- **Abdominal examination:** Abrasions, contusions, ecchymosis may indicate underlying visceral damage.

1. **London's sign:** Bruising of abdominal wall underlying hollow visceral perforation.
2. **Seat belt sign: Contusion across lower abdomen.**
3. **Saree sign** (Fig. 36.28): Across upper abdomen may be associated with pancreatic/bowel injuries.
4. **Cullen's sign:** Ecchymosis in the umbilicus
5. **Grey Turner's sign:** Ecchymosis in the flanks
6. **Rib fractures on right side:** Look for liver injury
7. **Rib fractures on the left side:** Look for splenic injury
8. Auscultation of **bowel sounds in the thorax**: Diaphragmatic injury
9. **Rectal examination:** Floating prostate, bony penetration—pelvic fracture, high riding prostate.
   - **Vermooten's sign:** Urethral injury—in cases of rupture of membranous part of urethra, prostate will not be palpable as it is displaced upwards. It is called **high riding prostate**.

Tertiary survey

Repeat primary survey, secondary survey and repeat laboratory/imaging studies (*see* Key Box 36.3 for wisdom lines in blunt abdominal trauma).

KEY BOX 36.3

BLUNT ABDOMINAL TRAUMA: WISDOM LINES

1. First priority in BAT is to assess and manage airway, breathing, with cervical spine precautions and circulation.
2. Immediate **splinting of fractures** and control haemorrhage.
3. Resuscitation using wide bore cannulae—'2 litres of fluid'
   If a hypotensive patient responds to fast transfusion of warm crystalloid solution, it means it is a hypovolaemic shock. It rules out head injury.
4. FAST / CT scan and decide—unstable patient will be taken to surgery immediately.
5. Frequent, repeated examination—any new signs in a stable patient such as
   - Tachycardia, Hypotension } Continuing haemorrhage
   - Temperature, Oliguria, Jaundice } Early sepsis, intestinal gangrene or perforation
   - Tachypnoea
   - Hazy chest X-ray, Nasogastric bleeding } Septic shock or diaphragmatic injuries
   - Immediate laparotomy is indicated

LIVER INJURIES

Liver injury should be suspected when a patient with suspected blunt injury abdomen is brought with the following features:

- Right lower ribs fracture
- Injury marks on the lower chest or upper abdomen.
- Patient with persistent hypotension or patient who had shock following blunt injury abdomen.
- A child can have liver injury without fracture of ribs because of elastic nature of the rib cage.

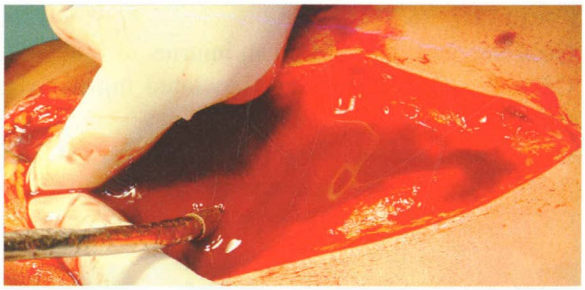


Fig. 36.5: Massive bleeding—challenging task (*Courtesy:* Dr Suni Krishna, Assistant Professor, KMC, Manipal)

Clinical presentation

- The most common presentation is features of intraperitonea haemorrhage, which includes hypotension, thready pulse abdominal distension as in Fig. 36.5. Peritoneal signs are minimal as early bleeding does not produce much peritonea irritation.
- However, massive lacerations of the liver including stellate fractures present with rapidly developing hypotension and shock, which are life-threatening.

Investigations

- Ultrasonography and more precisely CT scan should be done in all patients who are haemodynamically stable with or without support (Figs 36.6 to 36.9 and Key Box 36.4).
- Also *see* Table 36.1

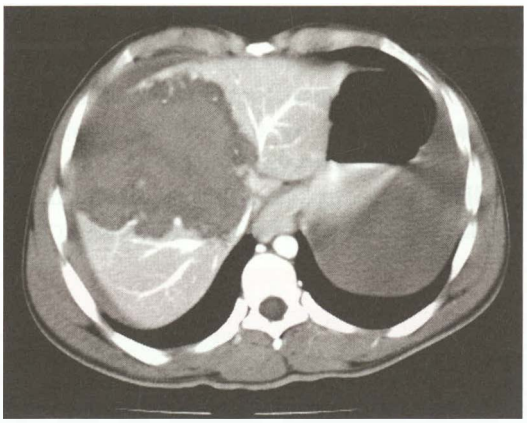
KEY BOX 36.4

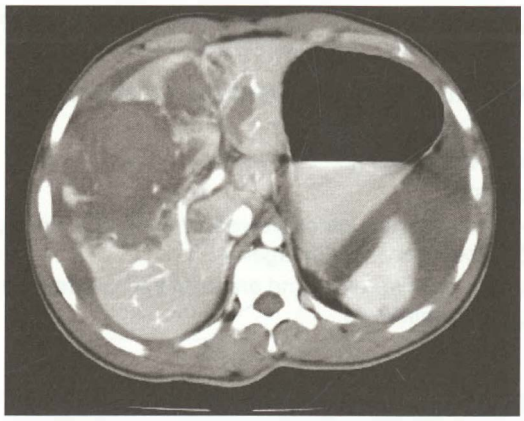
CT SCAN WITH IV CONTRAST

- It can grade the liver injury
- It can guide a conservative or operative treatment.
- It also rules out other injuries
- Grade I and Grade II injuries can be managed by non-operative treatment.
- Free contrast in and around the liver is indicative of active bleeding.

Table 36.1 Liver injury scale

Grade	Injury	Description
I	Haematoma	Subcapsular haematoma; < 10% surface area
	Laceration	Capsular tear, < 1 cm parenchymal depth
II	Haematoma diameter	Subcapsular, 10–50% surface area; intraparenchymal extension < 10 cm
	Laceration	< 10 cm long; 1–3 cm parenchymal depth
III	Haematoma	Subcapsular, > 50% surface area; expanding intraparenchymal haematoma of > 10 cm or expanding
	Laceration	> 3 cm, intraparenchymal depth
IV	Laceration	Parenchymal disruption of 1–3 Couinaud's segments within a single lobe
V	Laceration	Parenchymal disruption > 3 Couinaud's segments within a single lobe
	Vascular	Retrohepatic vena cava/central major hepatic veins
VI	Vascular	Hepatic avulsion





Figs 36.6 and 36.7: CECT arterial phase—Grade IV laceration of the liver with haemoperitoneum—interestingly patient was stable, managed conservatively, got discharged on 10th day

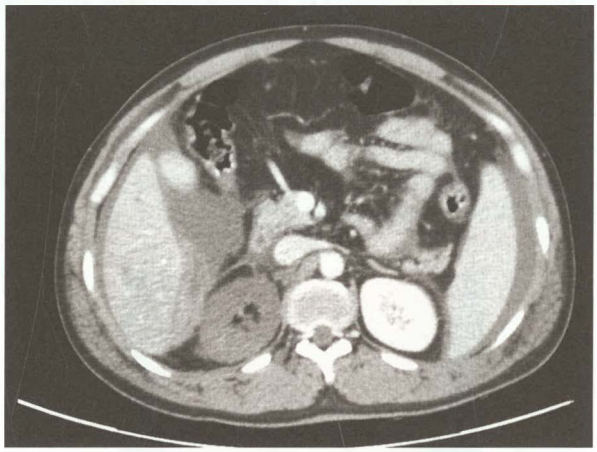


Fig. 36.8: Coronal section liver and kidney injury

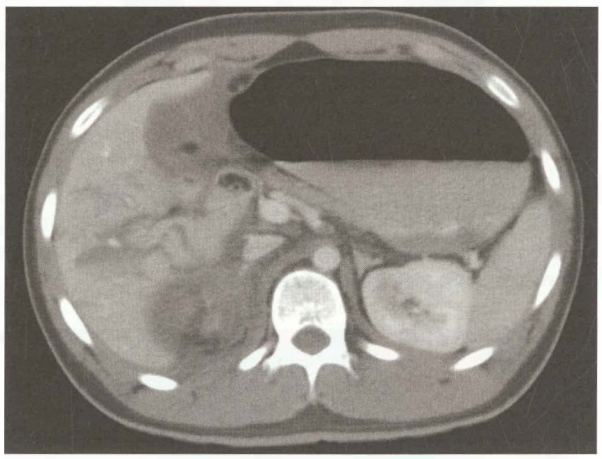


Fig. 36.9: Liver injury Grade II

Treatment (Key Boxes 36.5 and 36.6)

1. **Simple lacerations which are not bleeding at laparotomy:** A drain is kept in the liver bed, blood and clots are sucked out and peritoneal wash is given.
2. **Simple laceration with bleeding:** It is sutured by interlocking horizontal mattress sutures by using special liver suturing needle. If too much tension is applied while suturing, cutting through can occur. Omentum can be used as a **Plug** in between the laceration (Fig. 36.10). Absorbable sutures are used.

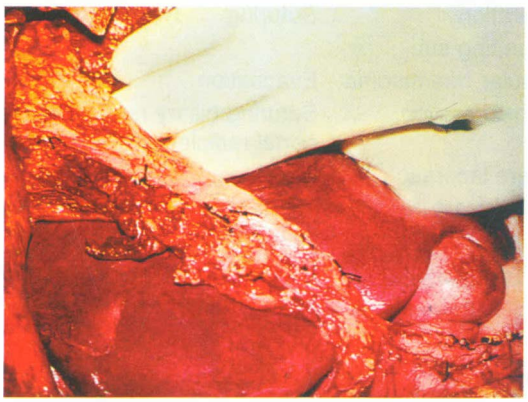


Fig. 36.10: Liver injury—use of omentum

3. **Subcapsular haematoma:** If present, should be evacuated.
4. **Deep laceration with bleeding:** In such situations, wound should be opened. Dead liver parenchyma is removed, bleeding vessel at depth and biliary radicle are ligated. It is described as **tractotomy**.
5. **Severe lacerations:** These injuries present with massive bleeding. Temporary control is obtained by compression of portal vein and hepatic artery in gastrohepatic omentum in front of **foramen of Winslow (Pringle manoeuvre)**. If bleeding stops, portal veins or branches of hepatic artery are damaged. If bleeding continues, hepatic veins are the source of bleeding. Visualisation of source of bleeding with debridement of avascular liver tissue is done by finger fracture method. **Perihepatic packing can be used to compress the liver as a temporary measure to buy time for resuscitation, to explore rest of the abdomen or as a definitive treatment when other measures fail.** Pack is usually removed after 24–48 hours (Fig. 36.11).
   - Nonanatomical resection may have to be done, in a few cases.

KEY BOX 36.5

HAEMOSTATIC TECHNIQUES AT SURGERY

- Chromic catgut liver suture
- Perihepatic packing
- Resection
- Argon beam coagulator followed by fibrin glue and sheet of Surgicel (haemostatic agent)
- Selective arteriography and embolisation in arteriovenous fistula or haemobilia.

KEY BOX 36.6

SUMMARY OF MANAGEMENT OF LIVER INJURIES

• Laceration	Suturing
• Expanding sub-capsular haematoma	Evacuation
• Deep laceration	Suturing biliary radicles and portal radicles, packing, etc.
• Severe laceration	Debridement
• Stellate fracture	Tractotomy; hepatic artery ligation
• Complex injuries	Ligation of hepatic vein, portal vein branches or lobectomy, etc.

6. **Complex liver injuries:** These injuries involve hepatic veins, retrohepatic vena cava or branches of portal vein resulting in massive haemorrhage. This type of massive injury can be managed by a large thoracoabdominal incision or abdominosternal incision by doing sternotomy. Division of the right triangular ligament helps in visualising bleeding from hepatic veins.

- **Schrock shunt:** Failure of Pringle manoeuvre means juxtahepatic and retrohepatic vena caval injuries. In such cases, Heaney manoeuvre clamping both infra and suprahepatic vena cava followed by atriocaval shunt or venovenous bypass can be done.

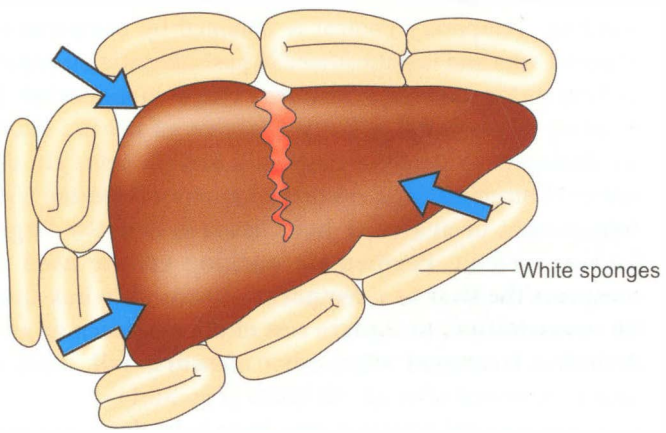


Fig. 36.11: Perihepatic packing

Complications of liver injuries

1. Massive bleeding, hypovolaemia and cardiac arrest.
2. Haematoma can get infected resulting in an **abscess**.
3. Haematoma can rupture into the peritoneal cavity resultin in leakage of bile—biliary peritonitis.
4. **Haemobilia** refers to rupture of the haematoma into the bil duct—it may result in massive haematemesis or melaena.

CLINICAL NOTES

A 24-year-old male patient was operated for blunt injury abdomen by laparotomy. Liver laceration was found. It was sutured.

After 15 days, he came to our hospital with haematemesis. Initially it was diagnosed as erosive gastritis. Ultrasonography revealed a pseudoaneurysm of one of the branches of middle hepatic artery. CT angiography and embolisation was tried but not successful. He had another bout of haematemesis. He underwent exploratory laparotomy and ligation of branches of middle hepatic artery. Bleeding stopped. Now since 6 months, no further attacks of haematemesis (Fig. 36.12). *It was a case of* ***haemobilia*** (Fig. 36.13).

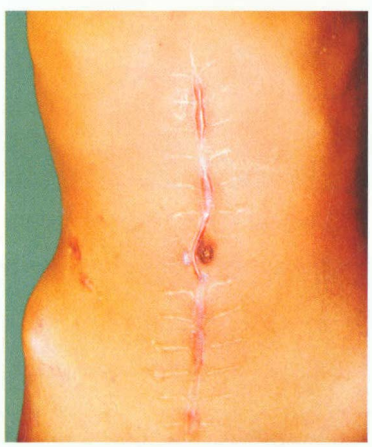


Fig. 36.12: Laparotomy scar of the patient who had liver laceration which was sutured. He presented with massive haematemesis later—**a case of haemobilia**

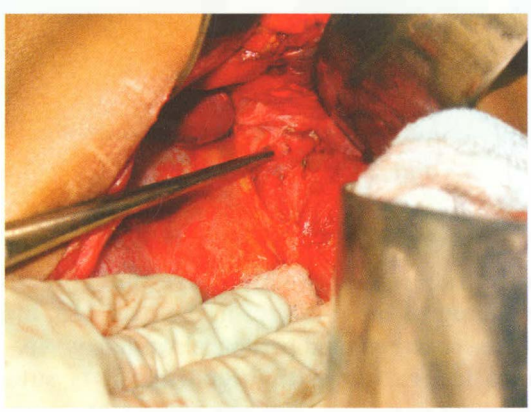


Fig. 36.13: Branch of middle hepatic artery ligated

SMALL BOWEL INJURIES

- The shearing injuries produce either disruption or laceration of the bowel between fixed and mobile points, i.e. at the **duodenojejunal flexure or at ileocaecal junction**. These are the most common sites of small bowel injuries.
- Injury to the small bowel can also occur due to crush injury between spine and a steering wheel or handle bars, etc.
- Bruising on the abdominal wall may suggest perforation (Fig. 36.14).
- Mesentery and its vessels also get damaged and bleeding can be sufficient to produce hypovolaemia and shock (Fig. 36.15).

Fig. 36.14: Observe London's sign: Bruising (imprint abrasion) over the abdominal wall signifying hollow viscus perforation

Fig. 36.15: Gangrene of the jejunal loop due to injury to mesenteric blood vessels in the same patient in Fig. 36.14

Clinical presentation

1. **Acute abdominal pain:** Features are like that of any perforation peritonitis with guarding and rigidity. Erect abdominal X-ray shows gas under the diaphragm.
2. **Features of peritonitis** with haemoperitoneum is the result of bowel injury with bleeding from the mesentery.
3. **Occult or hidden perforation:** A small perforation gets sealed off by coils of bowel and omentum. Most of these patients present with abdominal pain. However, very often, features of peritonitis are missed as a result of other associated injuries such as fracture pelvis or retroperitoneal haematoma. After 3–4 days, a localised abscess may form and rupture into the peritoneal cavity, resulting in peritonitis. This is aggravated by intake of oral fluids which stimulate peristalsis. **Repeated examination is the most honoured, most fruitful investigation in blunt injuries of the abdomen** (*see* clinical notes).

Investigation

- X-ray abdomen, erect or lateral decubitus (Fig. 36.16) demonstrates free gas under the right dome of the diaphragm in majority of cases. Four quadrant tap or diagnostic peritoneal lavage is also useful.
- When in doubt, CT scan of abdomen should be requested to diagnose hollow viscus perforation and bleeding.

CLINICAL NOTES

A 23-year-old male with fracture femur and pelvic fracture was admitted to the hospital after 12 hours of injury. A general surgeon was consulted to rule out an intra-abdominal injury. Pulse rate was 100/min and on deep palpation, there was tenderness in the right iliac fossa. Keeping in mind associated pelvic injury, it was decided to treat him conservatively. X-ray abdomen left lateral decubitus (Fig. 36.16) *(erect film could not be taken as patient could not stand) film did not show free intraperitoneal air (gas). Ultrasound revealed a retroperitoneal haematoma of 8 cm × 3 cm.*

The patient was treated conservatively with Ryle's tube for 3 days. On the 4th day, oral fluids were started, as patient passed stools once. On 7th day morning, patient had tachypnoea. Pulse was 120/min, BP was 90/60 mm of Hg. Previous 24 hours urine output was only 450 ml. Abdominal examination revealed guarding and rigidity in the right iliac fossa. It was decided to do a laparotomy. At laparotomy, there was a small 2 cm perforation in the ileum with bilioma surrounded by intestinal loops and gross contamination of peritoneal cavity. The perforation was closed and the peritoneal cavity was drained. The patient made a good recovery from septicaemia, thanks to early antibiotics and surgery.

PHOTOGRAPHS OF SMALL INTESTINAL PERFORATION (Figs 36.17 to 36.21)

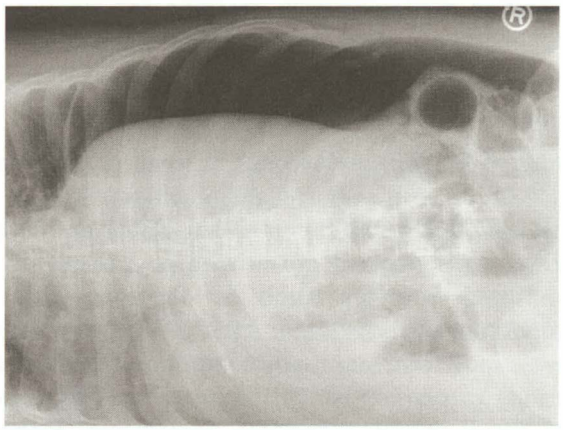


Fig. 36.16: Lateral decubitus picture is extremely useful in polytrauma cases when patient is unable to stand due to fractured limbs. However, **CT scan is the best investigation in blunt abdominal trauma** because it not only detects pneumoperitoneum but also other injuries

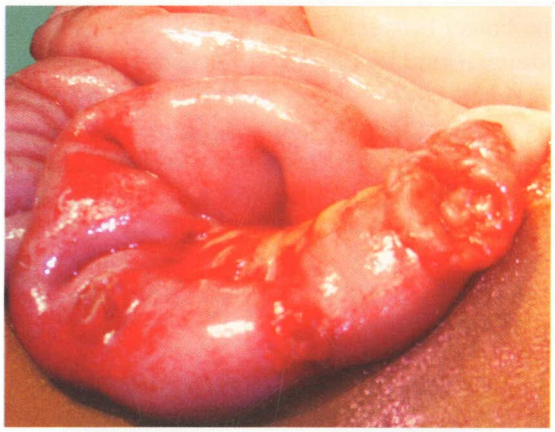


Fig. 36.17: Jejunal transection—6 cm away from DJ flexure. It is better to resect and anastomose in this type of cases (*Courtesy*: Dr Saurabh Aggarwal, Associate Prof, KMC, Manipal)

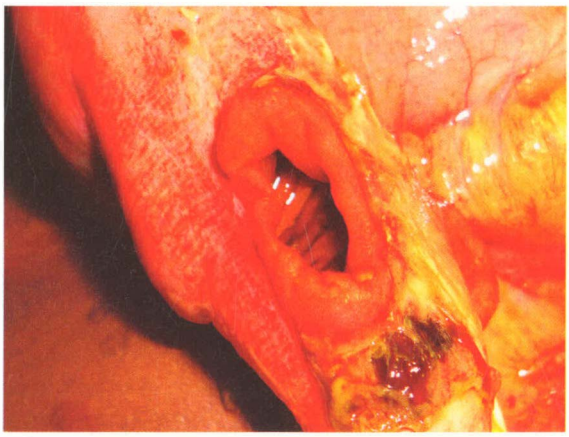


Fig. 36.18: 72 hours old perforation close to ileocaecal junction. Limited colectomy done

Fig. 36.19: Anastomotic leak following limited colectomy. Ileostomy done. It was closed after 8 weeks. You can see the wound infection

Fig. 36.20: Food particles—definite indication if found in DPL for laparotomy

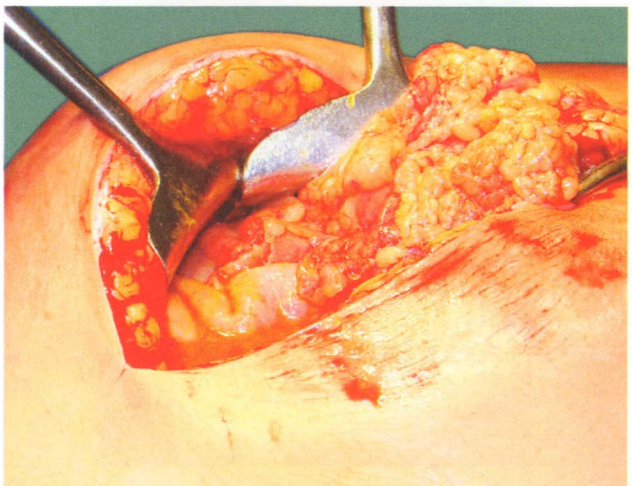


Fig. 36.21: Bile and blood in the peritoneal cavity

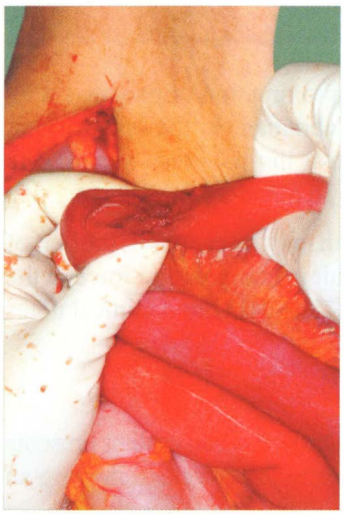


Fig. 36.22: Peritonitis following ileal perforation. Golden time to operate is within 6 hours of perforation

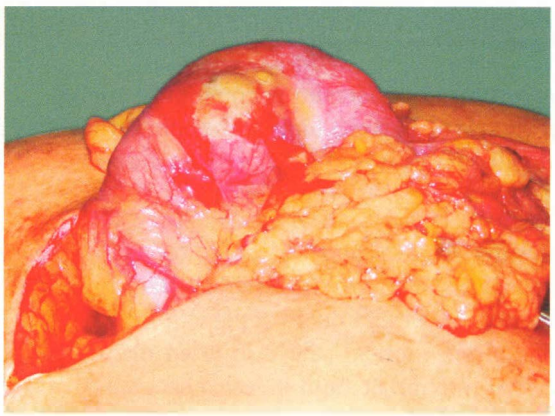


Fig. 36.23: Colonic perforations. The patient presented 7 days later with abdominal distension and early sepsis—steering wheel injury to the transverse colon

Treatment

Golden time to operate is within 6 hours

- **Perforation:** Single or multiple, have to be closed, after trimming the edges by using nonabsorbable sutures such as silk.
- A **lacerated** or a macerated bowel has to be resected.
- Bleeding mesenteric vessels have to be ligated, haematoma must be evacuated and bowel should be inspected for any ischaemia. Food particles and bile should be evacuated (Figs 36.17 and 36.23).
- A perforation of ileum **close to the ileocaecal junction** is treated by **ileocolectomy** rather than simple closure for the fear of enterocutaneous fistula, due to suture line leakage.

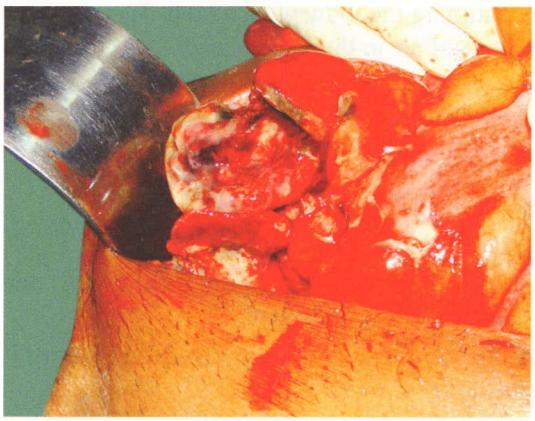


Fig. 36.24: Sigmoid perforation

KEY BOX 36.7

SMALL BOWEL INJURY

- Acute abdominal pain
- Guarding and rigidity
- Rebound tenderness
- Gas under diaphragm in plain X-ray abdomen erect
- Laparotomy and closure of perforation

COLONIC INJURIES

- Blunt injury of the colon is not uncommon.
- Mobile sigmoid is more prone for injury than fixed parts.
- Steering wheel injury can directly crush the transverse colon and can cause perforation.
- Bruise or laceration of the colon can undergo ischaemic necrosis and it can present after 5–7 days with signs of peritonitis/sepsis (Figs 36.23 and 36.25).
- Diagnosis is by clinical examination/contrast enhanced CT scan.
- Depending upon the contamination, contusion or laceration and duration of injury, treatment can be resection and anastomosis within 6–8 hours of the injury or simple suturing or diversion colostomy if gross contamination is present.
- Even in penetrating injury, primary closure can be done.

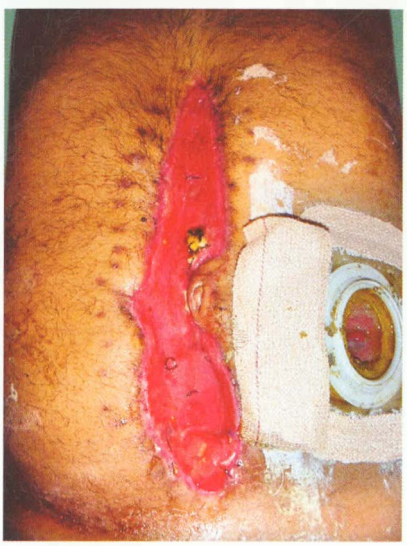


Fig. 36.25: Traumatic faecal fistula

DUODENAL INJURIES

- Retroperitoneal duodenum is commonly injured.
- Steering wheel, belt or a blow in the epigastrium may injure the duodenum as it is crushed against the spine.

Clinical features

- Peritonitis features are not common as it is the retroperitoneal duodenum (part II and III) that is injured.
- Tenderness is present on deep palpation.
- Being retroperitoneal, these injuries manifest late with abscess formation or fluid in lesser sac, etc.

Investigations

- X-ray abdomen
  - Obliteration of psoas shadow
  - Air outlining the kidney—Chilaiditi's sign (Fig. 36.26)
  - Absence of air in the duodenum
- Raised serum amylase is one of the biochemical parameters that should arouse a suspicion of pancreatic injuries along with duodenal injury.

Treatment

- Golden time to operate is within 6 hours.
- When in doubt, about narrowing of lumen, duodenojejunostomy may be indicated.
- When in doubt regarding duodenal fistula, tube duodenostomy is done.
- Duodenal haematoma is managed conservatively.

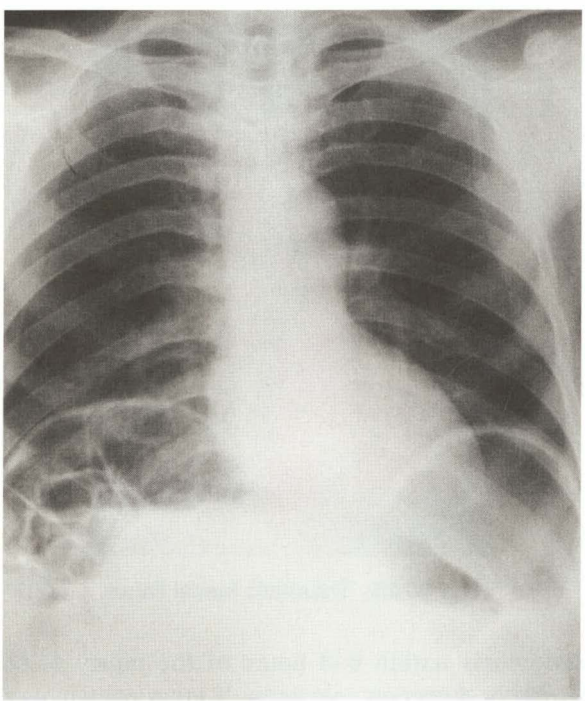


Fig. 36.26: Air outlining small portion of the kidney and air pockets under the diaphragm—Chilaiditi's sign

PANCREATIC INJURIES

Because of anatomical close approximation of pancreas with vertebral column, blunt injury abdomen in the epigastrium, kicks or **seat belt injuries** crush the pancreas against **the vertebral column** (Key Boxes 36.8 and 36.9).

Mill belt: Saree or *churidar* cloth may be caught in the belt of a running conveyor belt and result in compression force in the centre of the abdomen (Figs 36.27 to 36.31 and clinical notes *see* next page).

Diagnosis

- Pancreatic injury alone is diagnosed when patient presents with a pseudocyst of the pancreas 2–3 weeks following an injury.
- Very often, laparotomy is done for haemorrhage or perforation. In such situations, retroperitoneal bleeding, collection of bile or collection of fluid in the lesser sac arouses suspicion of pancreatic injuries.

Treatment

1. **Pseudocyst** following blunt injury abdomen invariably requires **surgical drainage,** e.g. **cystogastrostomy** because of injury to pancreatic duct.
2. Injury to body and tail require **subtotal pancreatectomy** with splenectomy.
3. Rarely, **pancreaticoduodenectomy** may be required for significant injury to the head of pancreas with injury to the duodenum.

Complications

- Pancreatic fistula
- Pancreatic pseudocyst
- Pleural effusion

KEY BOX 36.8

PANCREATIC INJURIES

- Anatomically hidden
- Very often, injuries missed
- Peritonitis features are not seen
- Dangerous because of enzymatic activation
- Can manifest as pleural effusion

KEY BOX 36.9

PANCREATICODUODENAL INJURIES

- Diagnosed late
- Peritonitis features are minimal
- Shock is very rare
- At laparotomy, they are missed
- Surgical treatment needs more skill and experience.
- Feeding jejunostomy is very useful
- Mortality and morbidity around 50%

CLINICAL NOTES

Indian women—vulnerability *(Figs 36.27 to 36.31)*
The saree and churidar are traditional Indian dresses. One end of the saree is tied around the waist and the other draped freely along the shoulder. If care is not taken, the free ***end often gets caught in the mill belt and causes*** *injuries* ***around the waist*** *(blunt abdominal trauma) due to the drag* ***created****. The impression created on the abdomen is by* ***the saree*** *and not the mill belt. The shawl of the churidar (has two free ends) can get caught in a belt or a wheel to cause injuries around the neck. The drag may be sufficient to cause even strangulation. The author remembers a case of scalp avulsion due to long hair of a woman getting caught in a mill belt.*

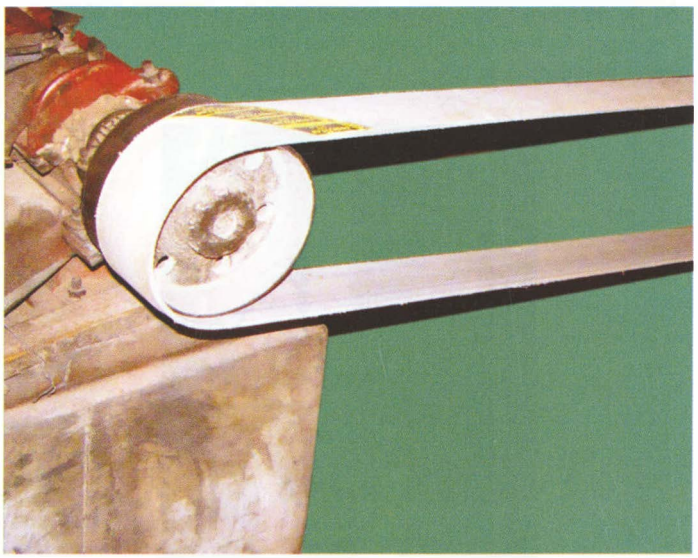


Fig. 36.27: Grinding mill having a conveyor belt (*Courtesy:* Dr Rakesh Hegde, Associate Professor, Department of Surgery, KMC, Manipal, 2006–2009)

Fig. 36.28: Impression of the saree all around waist in a lady who was dragged by the conveyor belt when her saree got caught

Fig. 36.29: In the OT

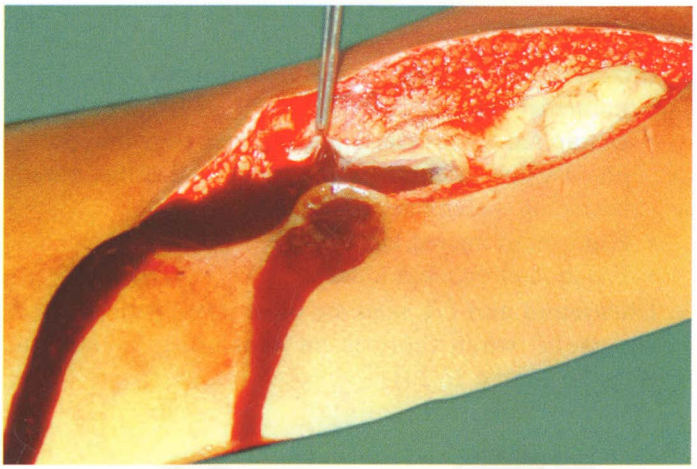


Fig. 36.30: On laparotomy, blood gushing out (*Courtesy:* Dr Raghunath Prabhu, Associate Prof, KMC, Manipal)

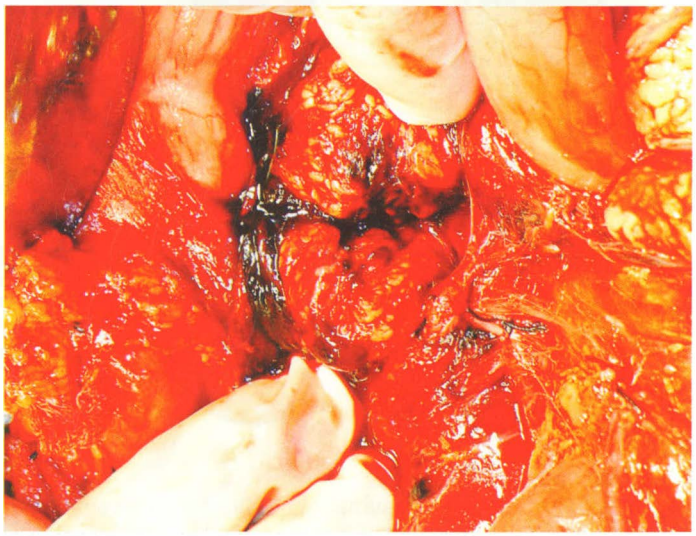


Fig. 36.31: Pancreas split into 2 parts

RENAL INJURIES

Types (Fig. 36.32)

I. **Minor injuries:** Subcapsular haematoma, minor laceration and renal contusions.
II. **Major injuries:** Bleeding into renal pelvis from laceration of medulla, corticomedullary rupture, hilar injury.

Clinical features

- **Haematuria** is the most important (80–90%) sign of renal injury. It may be mild, or sometimes can be massive depending upon the extent of injury. It may be absent in renal pedicle avulsion.
- **Loin bulge** due to perinephric haematoma
- **Bruising** of soft tissue in the loin
- Retroperitoneal haematoma compressing on splanchnic nerves (meteorism) results in paralytic ileus, which causes **abdominal distension.**
- Associated injuries such as fractures of the transverse process of lumbar spine may be present.

Investigations

1. **Intravenous pyelography can demonstrate**
   - Intrarenal extravasation
   - Extrarenal extravasation (pararenal pseudohydronephrosis due to extravasated blood and urine, slowly occluding pelviureteric junction).
   - Function of injured kidney
   - Function of opposite kidney
2. **Ultrasound and CT scan** are other investigations which are useful when there is an expanding haematoma (Figs 36.33 and 36.34).

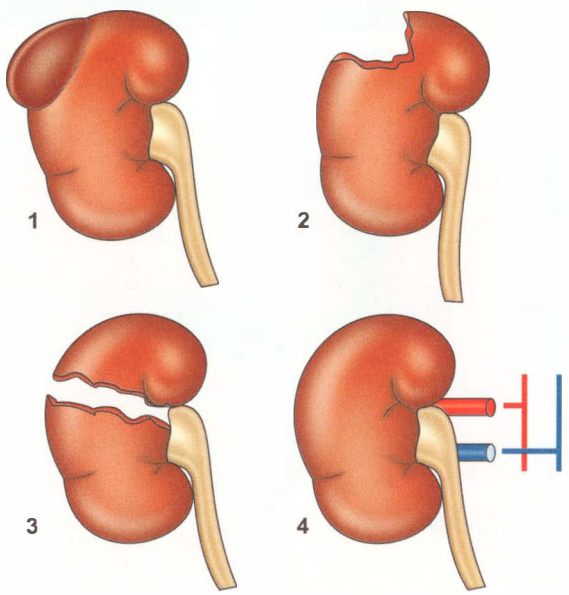


Fig. 36.32: Closed renal trauma: (1) Subcapsular haematoma, (2) laceration, (3) avulsion of one of the poles and (4) avulsion of renal pedicle

Fig. 36.33: Renal trauma: Right kidney is transected and upper pole is displaced by a large haematoma. Left kidney is normal (*Courtesy:* Dr Padmaraj Hedge, Prof and Head, Dept. of Urology, KMC, Manipal)

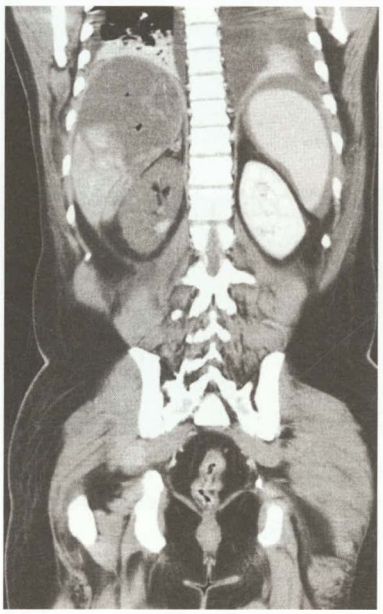


Fig. 36.34: Liver and kidney injury

Treatment

1. **Conservative:** Minor injuries are managed conservatively with close monitoring of vital signs such as pulse, blood pressure, temperature and respiration, Hb% and PCV.
   - Sedation and analgesics are also given
2. **Surgical exploration**
   - **Small laceration** sutured over gel foam or by using detached muscle.
   - **Major laceration** involving one pole—a partial nephrectomy is done.
   - **Major multiple lacerations**, avulsions, require nephrectomy.

RETROPERITONEAL HAEMATOMA

- It is quite common because of accidents, fall from height.
- Fracture vertebrae, fracture pelvis, injury to retroperitoneal veins.
- Bleeding from vena cava and aorta can be fatal.
- Haematomas which are not expanding should not be disturbed.

Diagnosis and management of retroperitoneal haematoma as shown in Fig. 36.35.

Pelvic fractures and retroperitoneal haematoma

- Pelvic fractures are also an important cause of retroperitoneal haematoma.
- The most frequent mechanisms causing pelvic fractures are motor vehicle accidents, motorcycle accidents, falls and accidents involving pedestrians. Associated injuries to urethra in males should be ruled out first. Per rectal examination should be done to evaluate the position of the prostate. CT scan is done to assess pelvic fracture and also to assess retroperitoneal haematoma (Figs 36.36 and 36.37).
- Retroperitoneal bleeding can be arterial, venous, or osseous in origin. Unstable pelvic fractures are generally associated with increased blood loss. Posterior fractures with involvement of the sacroiliac joint are frequently associated with arterial bleeding, which can be controlled by embolisation of the bleeding vessel, usually branches of the internal iliac artery. Unstable fractures should be fixed by external fixation. Expanding haematoma should be explored. Control the bleeders, otherwise pack the pelvis. Nonexpanding haematoma should not be explored. Wait and watch.

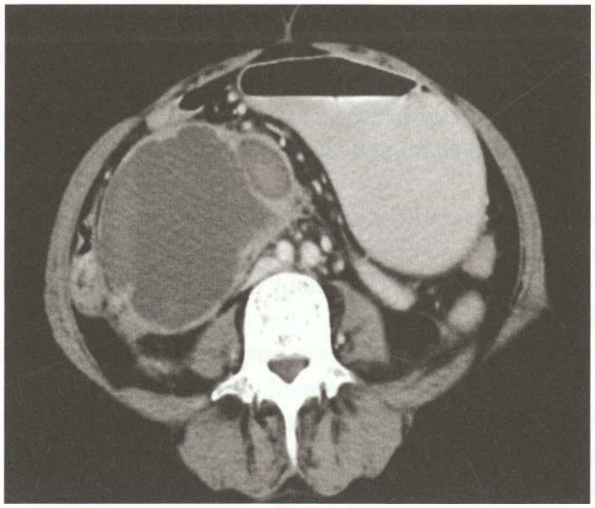


Fig. 36.36: Retroperitoneal haematoma contrast CT

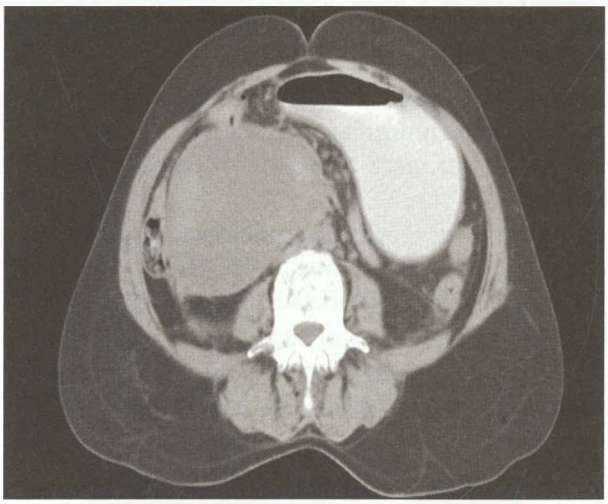


Fig. 36.37: Retroperitoneal haematoma

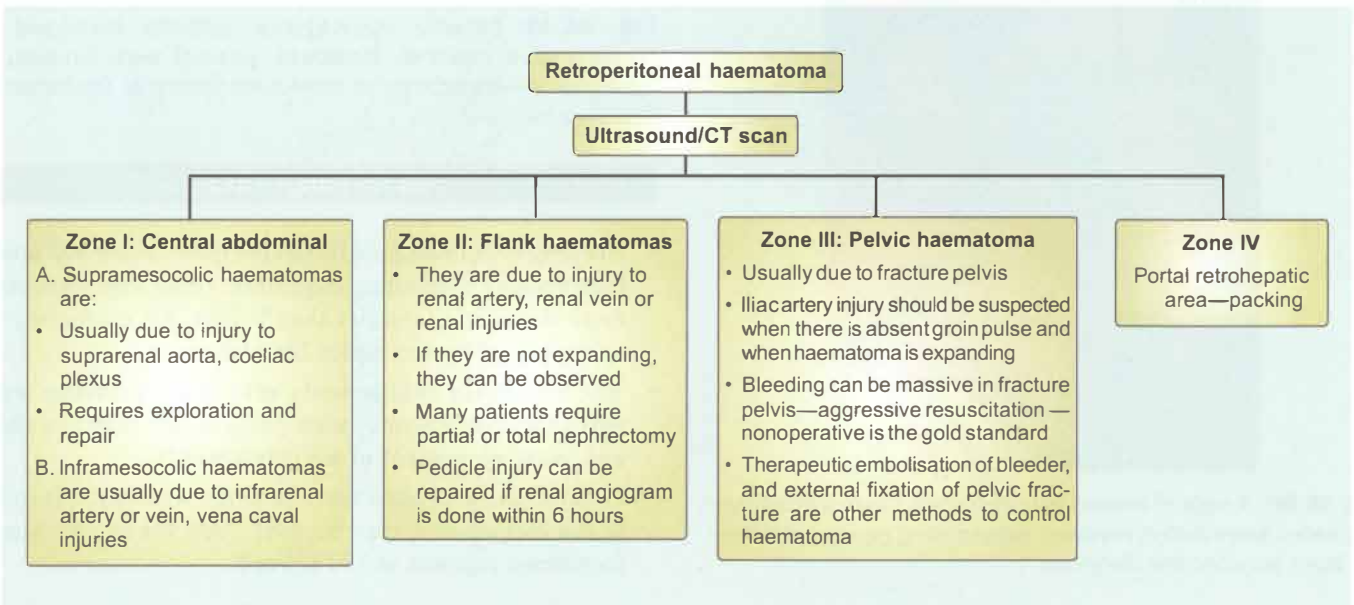


Fig. 36.35: Diagnosis and management of retroperitoneal haematoma

GENERAL PRINCIPLES IN A BLUNT INJURY ABDOMEN

- A patient should be admitted to the hospital and carefully monitored if there is a slight doubt regarding blunt injury abdomen.
- **Repeated examination,** careful monitoring of pulse rate, temperature and blood pressure, chest X-ray, estimation of Hb%, frequently help in many cases of silent blunt injuries (Fig. 36.38).
- Most of the cases today are **polytrauma** cases, Hence, all systems should be examined. Among all these, priority should be given to life-threatening, salvageable injuries such as **extradural haematoma, haemothorax, splenic injuries, liver injuries**.
- It is easier to make a diagnosis of fracture[1] (revealed injuries) which can be treated later. FRACTURE CAN WAIT BUT NOT RUPTURE. Concealed injuries should be carefully looked for.
- Undoubtedly, **diagnostic peritoneal lavage, ultrasound** (CT scan is the immediate noninvasive investigation) help in diagnosis of more than 90% of cases of blunt injury abdomen.
- Adequate blood, appropriate antibiotics, aggressive resuscitation before surgery to treat hypovolaemia and shock are the major factors which decide the outcome of surgery.
- In a major accident involving many patients and limited resources, quick decision should be taken regarding **triage**—who can be saved, who cannot be saved (Figs 36.39 and 36.40).

Fig. 36.38: A case of missed liver injury with haemoperitoneum. Repeated examination revealed deteriorating general condition. CT scan provided the diagnosis

Fig. 36.39: Pelvic fracture with ileal injury—frequent clinical examination revealed guarding and rigidity (*Courtesy:* Dr Rajesh Sisodia, Associate Prof, KMC, Manipal)

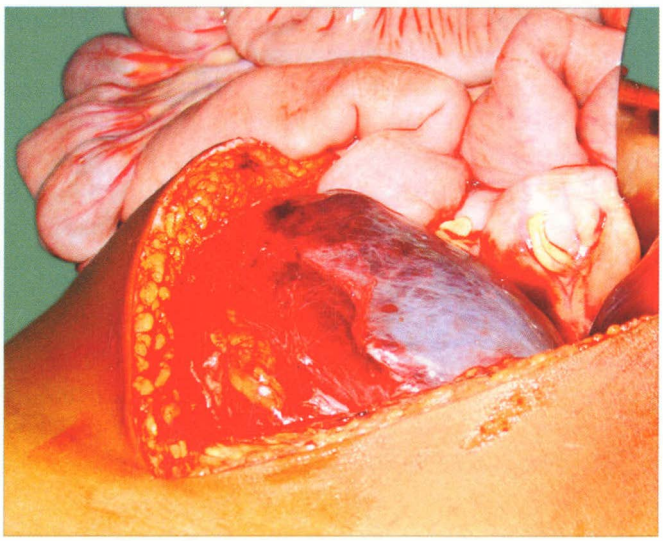


Fig. 36.40: Splenic haematoma—initially managed by conservative method. However, patient was developing hypotension—importance of careful monitoring of the patient in intensive care unit

BLAST INJURIES

- Bursting of bombs or shells rupture their casing and impart high velocity to resulting fragments. These fragments cause more devastating injuries than bullets. All explosives are accompanied by a complex blast wave.
- **The two main components are: Blast pressure wave** (dynamic overpressure) with positive and negative phase and mass **movement of air (blast wind).**
- Positive phase of blast wave lasts few milliseconds (close to the explosion it may be over 7000 kN/m^2) (tympanic membrane ruptures at 150 kN/m^2).

[1]Even a grandmother can diagnose fractures. However, it needs lot of experience and skills in diagnosing blunt injury abdomen.

- Like sound waves blast, pressure waves flow over and around an obstruction and affect persons sheltering behind a wall. The pressure affecting such a person is known as incident pressure (pressure at 90° to direction of travel of blast shock front).
- Person standing in front of a wall facing an explosion is subject to added effect of reflected pressure. Mass movement of air displaces air at supersonic speed. This disrupts environment, hurting debris and people. Blast wave under the water travels at great speed and to greater distance. Injuries tend to be complex and severe.

PEARLS OF WISDOM

Structures injured by primary blast wave are ears, lungs, heart and gastrointestinal system.

- Most will have combination of blunt, blast and thermal injuries. Deafness, lung contusion, capillary leakage and haemorrhage into alveoli and ARDS precipitated by over transfusion are the features. Perforation of the intestines and penetration injuries to the eye are the other features.
- Management consists of resuscitation in a well-equipped trauma unit, blood transfusions, intensive care monitoring, antibiotics and appropriate surgical procedures.

WARFARE INJURIES

Penetrating missile wounds, injuries from blast phenomena and burns are typical features of modern conventional war. The most common wounding agent in surviving casualties is a fragment wound and not a bullet wound as many erroneously believe. The aim in modern war is to incapacitate and **not to kill**. Hence, large number of surviving casualties are a major financial and logistic burden on a nation engaged in war.

Wound Ballistics and Mechanisms of Injury

Bullets fired from hand guns are propelled at low velocity, have low available energy and result in low velocity transfer wounds (100–500 J), whereas those from assault rifle have high velocity and have high available energy (2000–3000 J) and they cause high energy transfer wounds. **Low energy** transfer wounds leave injury confined to wound tract. **High energy** transfer wounds causes local laceration, crush injury and also cause remote injury from wound tract due to temporary cavitation phenomena.

Management

- Entrance and exit wounds do not indicate considerable damage that may have occurred to deeper structures.
- Resuscitate as per ATLS guidelines.
- Record the wounds in case sheets, take photographs if necessary.
- Under anaesthesia, excise skin around entry and exit wounds, give liberal longitudinal incision through skin and deep fascia, which allows proper visualisation of underlying structures.
- Debride (cut till healthy tissues are seen) all dead tissues—dead muscle does not bleed or contract, looks dusky.
- Identify neurovascular bundles and examine them.
- Dissect and mark injured nerves for possible future repair.
- Repair arteries and veins if injured
- Give thorough wash and let out all the dirt.
- Injured tendons are trimmed and tied for easy identification at future surgery.
- Fix bones by appropriate methods.
- Cover the wound with absorbable dressing.
- Appropriate antibiotics and injection tetanus toxoid are given.
- Amputation may be necessary if limb is grossly mutilated
- Delayed primary closure is done (4–6 days later) once the wound starts healing.

MISSILE WOUNDS OF ABDOMEN

- Every penetrating and perforating missile wound of the abdomen should be explored by laparotomy. A full midline incision from xiphisternum to pubis is recommended and it may be extended to thorax if necessary.
- The rest of the treatment depends on the nature of injury. Bleeding mesenteric vessels are ligated, injured small bowel is repaired by suturing or by resection and anastomosis. In colonic injuries, simple closure or closure with protective colostomy is necessary depending upon the nature of the colonic injury and contamination.
- Liver, splenic, pancreatic and renal injuries have been discussed in respective chapters.

PENETRATING TRAUMA OF THE ABDOMEN

Today all penetrating injuries of the abdomen need not be explored by laparotomy. Good physical examination of the patient, entry and exit point, and haemodynamic status of the patient followed by investigations such as ultrasonogram and CT scan will guide the decision for laparotomy.

KEY BOX 36.10

INDICATIONS FOR LAPAROTOMY

- Tenderness, guarding, rigidity
- Unexplained shock
- Evisceration of contents
- Positive investigations
  - Positive DPL
  - Gas under diaphragm
  - IVP, cystoscopy, cystogram
  - Ultrasonogram
  - CT scan

TRIAGE

- The term **triage** is derived from the French word ***trier***, meaning **to sort**. This is a method of sorting out injured patients, during mass casualties depending on the severity of injury. If you are the first on the scene, first priority is to get expert help (call up fire services, regional trauma centre, etc).
- Triage is a skilled activity by a Trauma team, wherein there is a leader (usually senior-most doctor/surgeon) and there are many assistants. The leader sorts out patients (people) depending upon their severity of injury.
- Each of the patients can be colour coded (a coloured flag is attached to them). Examples:
  1. **Immediate help is necessary—Red:** Otherwise the person will die in a few minutes if no treatment is offered, e.g. obstructed airway, tension pneumothorax.
  2. **Urgent help is necessary—Yellow:** These patients may die in 1–2 hour if no treatment is given. Examples are cases of massive bleeding and hypovolaemia.
  3. **Delayed—Green:** These patients can wait. Examples are minor fractures.
  4. **Expectant—Blue:** Very severe injury and unsalvageable. An attempt to treat them, may delay treatment to other patients who are salvageable.
  5. **Dead people are flagged—white or black**.

PEARLS OF WISDOM

The leader assesses the patient—flags and moves forward and it is his assistant who carries out the necessary resuscitation. The leader should not resuscitate any patient. There are others waiting for his expert help.

KEY BOX 36.11

FUNDAMENTAL STEPS

- Airway management
- Breathing
- Circulation
- Disability and assessment of level of consciousness by Glasgow Coma Scale.
- Exposure of the patient fully for thorough examination.
- Finger evaluation and tubes

A common scheme

1. Can the patient walk?

Yes—Delayed (green)

No—Check for breathing

2. Is the patient breathing?

No: Open the airway

Are they breathing (ventilating) him?

Yes—immediate (red)

No—DEAD (white)

Yes: Count or estimate respiratory rate (over 15 sec).

< 10 to > 30 per minute—immediate (red)

10–30 per minute—check the circulation.

3. Check the circulation

Pulse >120/min (capillary refill >2 s)—immediate (red)

Pulse < 120/min (capillary refill < 2 s)—urgent (yellow).

- If any regional trauma centre is well equipped and nearby one has to transport all the injured patients to hospital (scoop and run), where expert help is available. Meanwhile trauma centre can be alerted about the arrival of casualties. If expert help is far away, then one may have to treat the patients at the accident site (stay and play). Resuscitation is done as per ATLS guidelines.

KEY BOX 36.12

QUICK REMINDER MNEMONIC AT TREATMENT CENTRE

- **L**ift jaw
- **I**ntubation: Airway
- **F**allen tongue/foreign body to be checked
- **T**racheostomy/cricothyrotomy/oropharyngeal or naso-pharyngeal throat suction
- **J**aw fractures to be ruled out

Remember as **LIFTJAW**

- During these exercises do not forget to take care of your own safety, in burning vehicles, burning or falling buildings, etc.

PEARLS OF WISDOM

An initial quick evaluation of the patient for anaemia, level of consciousness, if necessary volume replacement, application of cervical (C) collar to the neck, etc. are done.

Management (Key Boxes 36.11 and 36.12)

These six initial steps are included under primary survey and resuscitation in ATLS (advanced trauma life support) are given below.

Airway with cervical spine protection

- Rapid assessment of signs of obstruction—foreign body, laryngeal and faciomaxillary fracture, fallen back tongue.
- Lift the jaw, introduce airway, good suction of throat, intubation or even tracheostomy if necessary.
- In polytrauma, assume cervical spine fracture and cervical collar is applied immediately (Fig. 36.41).

Breathing and O_2 administration

- Rule out tension pneumothorax, multiple fractures of ribs and haemothorax, surgical emphysema, etc.
- All trauma patients should receive ***high-flow oxygen.***

Fig. 36.41: Management of a polytrauma patient in an intensive care unit—cervical collar has been applied and patient is being ventilated

PEARLS OF WISDOM

Tension pneumothorax is a clinical diagnosis. Do not delay the treatment while waiting for chest X-ray.

Circulation and control of bleeding

- Immediate and quick assessment of circulation status, evidence of shock and evidence of internal or external bleeding should be looked for.
- Feeble thready pulse, hypotension indicates volume or blood loss. Splenic, liver or mesenteric injuries can be diagnosed by ultrasound. Bleeding into the pleural space or pericardium needs further tests—CT scan or echo cardiography, etc.
- Assess the class of haemorrhage
- Two large bore intravenous lines to be placed.
- Initial bolus of 2 litres of crystalloid solutions.

PEARLS OF WISDOM

The immediate goal is to arrest bleeding rather than replacing the blood.

KEY BOX 36.13

DISABILITY: AVPU SYSTEM

- **A**wake
- Open eyes to **V**oice
- Open eyes to **P**ainful stimulus
- **U**narousable

Disability

Glasgow Coma Score and assessment of pupils can help immediately to assess the status of the patient.

Exposure with control of environment

Complete inspection of the patient by taking out clothing. Keep patient warm.

Fingers and tubes

- Quick examination of all orifices. Examples: Look for bleeding from the ear, nose, oral cavity, rectum, vagina, and urethra.
- Nasogastric tube, endotracheal tube, catheter, intravenous line or central line are the immediate requirements of a polytrauma patient.

Please note

Remember following mnemonics in blunt injury abdomen:

- AMPLE – page 887
- ABCDE – page 887
- FAST – page 886
- LIFT JAW – page 900
- AVPU – page 901

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All investigations have been updated.
- Liver and pancreatic injury have been discussed in more detail.
- More colour photographs and key boxes have been included.

MULTIPLE CHOICE QUESTIONS

1. The most common bedside investigation done for suspected blunt abdominal trauma for bleeding is:

A. CT scan
B. MRI scan
C. Diagnostic peritoneal lavage
D. Ultrasound

2. Which one of the followings is a definite indication for laparotomy in blunt injury abdomen?

A. Splenic injury
B. Pancreatic injury
C. Liver injury
D. Aspiration of bile in the peritoneal aspirate

3. If air bubble like picture is found within the thorax following blunt injury abdomen what do you suspect?

A. Splenic rupture
B. Liver injury
C. Injury to the stomach
D. Diaphragmatic injury

4. How do you rule out a head injury with factors given below?

A. Hypotension responding to fluid
B. CSF rhinorrhoea
C. Fracture skull
D. Hypertension and bradycardia

5. Which is an important sign of hollow viscus perforation?

A. Cullen's sign
B. Grey Turner's sign
C. Mallet Guy sign
D. London sign

6. The following are true for conservative management of liver injury *except*:

A. Hollow viscus injury should not be there
B. Free contrast in and around liver in CT scan
C. Grade I and Grade II injury
D. Haemodynamically stable patient

7. Pringle manoeuvre refers to:

A. Compression of left gastric artery to stop the bleeding from giant gastric ulcer
B. Compression of hepatic artery to stop the bleeding during liver resection
C. Compression of hepatic artery and portal vein in front of foramen of Winslow
D. Compression of gastroduodenal artery during Whipple's procedure

8. The salvage procedure to buy time in massive bleeding from liver include following:

A. Pringle manoeuvre
B. Plug by omentum
C. Perihepatic packing
D. Portovenous shunt

9. Perforation within 4 cm of the ileocaecal junction following blunt injury is better treated by:

A. Suturing and drainage
B. Resection and anstomosis and drainage
C. Suture and bypass
D. Exteriorisation

10. Which of the following is not a feature of retro-peritoneal duodenal perforation?

A. Can occur with steering wheel injury
B. Chilaiditi sign may be present
C. Free gas under diaphragm
D. Guarding and rigidity is minimal

ANSWERS

1 D	2 D	3 D	4 D	5 D	6 B	7 C	8 C	9 B	10 C

37

Abdominal Mass

- Clinical examination of abdominal mass
- Mass in the right iliac fossa
- Firm to hard nodular mass in the umbilical region
- The cystic mass in the abdomen
- Mass in the epigastrium
- Mass in the right hypochondrium
- What is new?/Recent advances

Introduction

The abdomen is like Pandora's box. However, a student who is examining a case of abdomen is like an investigating CBI officer. He has to collect information at every level of examination, i.e. history, past history, general examination and abdominal examination. An attempt has been done here to highlight the importance of history and clinical examination. **Ten points** in the history, if taken and analysed properly may give a definite **clue** in majority of cases. After getting this **clue**, clinical examination of the mass may become easy.

CLINICAL EXAMINATION OF ABDOMINAL MASS (CLINICS)

REGIONS IN THE ABDOMEN

- Abdomen is divided into nine regions (quadrants) by two horizontal lines and two vertical lines.
- Upper horizontal line or transpyloric line is midway between xiphisternum and umbilicus.
- **Lower hoizontal line (transtubercular line)** is the line joining iliac crest tubercles of each side, about 5 cm behind anterior superior iliac spine.
- The vertical lines are drawn on either side through midpoint between anterior superior iliac spine and symphysis pubis. Following are the nine regions of the abdomen (Fig. 37.1).
  1. Right hypochondrium
  2. Epigastrium
  3. Left hypochondrium
  4. Right lumbar region
  5. Umbilical region
  6. Left lumbar region
  7. Right iliac fossa
  8. Hypogastrium
  9. Left iliac fossa

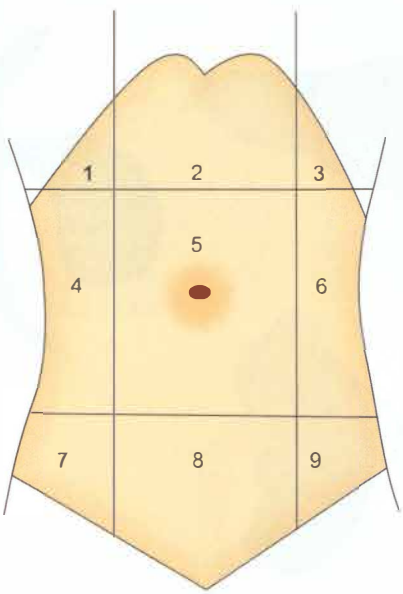


Fig. 37.1: Regions of the abdomen (*see* text)

HISTORY

1. Abdominal pain: It is present in most of the cases of abdominal mass. Abdominal pain can be of the following types:

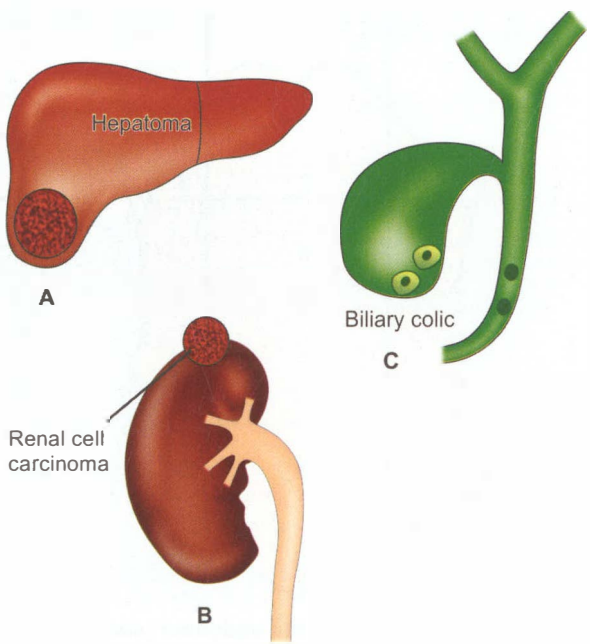
A. Dull aching pain: It suggests a solid organ enlargement. It is a continuous pain felt in the anatomical location of the swelling. Patients often describe it as a discomfort rather than pain.

Examples

- Liver enlargement: Pain in the right hypochondrium. It occurs due to stretching of **parietal capsule** (Glisson's) (Fig. 37.2A)
- Splenic enlargement: Pain in the left hypochondrium
- Renal enlargement: Pain in the back and costal region or costovertebral pain (Fig. 37.2B)
- Enlarged lymph nodes (para-aortic), pancreatic tumours: Backache

B. Colicky pain suggests hollow viscus obstruction. This pain is due to hyperperistalsis. It is severe and intermittent (comes and goes). Each attack may last for 5–10 minutes. The patient bends on himself, holds the abdomen and puts pressure on the abdomen which gives some kind of relief. Being visceral type of pain, it is not very well localised. Following are a few examples:

- Mass in the right iliac fossa (carcinoma caecum or ileocaecal tuberculosis). Initially there may be a vague discomfort. However, when partial obstruction occurs, it results in a colicky abdominal pain which is centrally located and sometimes unbearable.
- Ureteric colic and biliary colic (Fig. 37.2C).



Figs 37.2A to C: Source of the pain

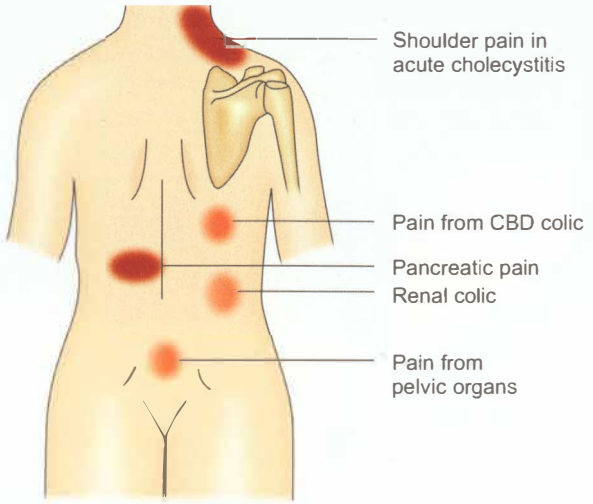


Fig. 37.3: Referred pain (posterior view)

- Carcinoma pyloric antrum or pyloric stenosis produces colicky upper abdominal pain with gastric peristalsis. However, this type of pain is not an unbearable one.

C. Referred pain: Tuberculosis of spine is a common problem in India. Often patients present with iliopsoas abscess. Patients can complain of referred pain in the lower abdomen (Fig. 37.3).

2. Sensation of fullness/early satiety

- Carcinoma of the stomach and pyloric obstruction. Also hepatoma or large pancreatic tumours can cause extraluminal compression on the stomach resulting in sensation of fullness in the abdomen.
- Early satiety is due to loss of receptive relaxation of stomach due to malignant infiltration of muscle layer.

3. Vomiting

- Persistent, profuse, projectile and nonbilious vomiting suggests pyloric stenosis. Chronic duodenal ulcer and carcinoma stomach are the common causes of pyloric obstruction (pain is absent/negligible).
- Persistent, profuse, projectile, bilious vomiting—intestinal obstruction. For example, ileocaecal tuberculosis, stricture of the small bowel, adhesions (pain is severe and colicky).

4. Haematemesis

- Epigastric mass suggests carcinoma stomach
- Splenomegaly may be an indication of portal hypertension.

5. Bleeding *per* rectum

- Fresh blood with or without melaena---carcinoma rectum
- Melaena—carcinoma stomach, portal hypertension

6. Loss of appetite and loss of weight

- These are common symptoms of GI malignancies. Please note that these two symptoms are seen not only in intra-abdominal malignancies but also in many diseases such

as tuberculosis. However, it should be noted that one of the **earliest signs of carcinoma stomach is loss of appetite. Severe weight loss is an early and important feature of carcinoma body of the pancreas.**

- Significant weight loss refers to loss of 10 kg or more in the last 6 months.

7. Bowel habits

- Fresh bleeding *per* rectum: Carcinoma rectum
- Blood and mucus (bloody slime): Carcinoma rectum
- Alternate constipation and diarrhoea: Carcinoma colon

8. Jaundice

- Progressive, persistent, pruritic jaundice: Periampullary carcinoma or carcinoma head of pancreas. However, in periampullary carcinoma, fluctuation can occur if growth ulcerates.
- Mild recurrent jaundice: Haemolytic anaemia.
- Intermittent jaundice, pain, fever: **Charcot's triad**—stone in the common bile duct.

9. Haematuria: Fresh bleeding/clots: Renal cell carcinoma.

10. Fever

- **High-grade fever**, with chills and rigors: Stone in common bile duct
- **Low-grade fever**: Hepatoma, renal cell carcinoma, lymphoma. Fever is due to some pyrogens released into circulation or due to tumour necrosis.
- In a **tropical country** like India, **hepatomas with fever** are often diagnosed as **amoebic liver abscess and mistreated.**

ON EXAMINATION

Inspection

- The patient is asked to breathe well with mouth open.
- Students should spend a few minutes watching the abdomen carefully.

1. Shape of the abdomen

- **Scaphoid** in normal cases
- **Protuberant** in fatty abdomen.
- Generalised distension with **fullness in the flanks** is usually due to **ascites**.
- Localised distension can be due to a mass
- Presence of **step ladder peristalsis** indicates **small bowel obstruction,** visible gastric **peristalsis indicates pyloric stenosis** and **right to left peristalsis** indicates **colonic obstruction.**

2. Restricted movement of any one region of the abdomen indicates an inflammatory pathology.

3. Umbilical nodule (Sister Joseph's) indicates intra-abdominal malignancy (carcinoma of stomach, colon, pancreas)

4. Details about the mass such as size, shape, surface, borders, movement with respiration have to be mentioned if mass is visible. If the details about the mass cannot be appreciated or if mass is not clear on inspection, **it is better to say "there is fullness"** rather than trying to manipulate the details about the mass.

5. Inspection of male genitalia: If scrotum is empty, it could be a case of undescended testis[1,2].

Palpation

Methods of palpation

Following are the methods of palpation available to the clinician and done depending upon the merits of the case:

1. **Superficial palpation:** Gentle superficial palpation of the abdomen gains confidence of the patient. It can detect superficial lesion of the abdominal wall such as lipomatosis, neurofibromas or fibromas, etc. It can also detect an area of tenderness, so that clinician is careful while doing deeper palpation. Superficial palpation is done with the flat of the hand or fingers.
2. **Deep palpation:** These are important requirements for deeper palpation:
   - Patient should be well-relaxed, with flexion of the knee for about 45 degrees.
   - The patient's face should be turned to the opposite side and he is asked to breathe comfortably with open mouth.
   - **Deep palpation** should be started from the **quadrant situated diagonally opposite** to the site of pain.
   - Palpation should **cover not only the 9 quadrants of the abdomen,** but also **2 more quadrants, i.e. the 2 renal angles** and **12th quadrant—external genitalia in males[3].**
   - Deep palpation is carried out with the palmar surface of the fingers and some degree of angulation depending upon the depth of palpation.

Tests

1. Movement with respiration: This test is done by placing the fingers (hand) over the lower border of the swelling and the patient is asked to take a deep breath. Movement with respiration is positive when there is "up and down" movements not anteroposterior movement. Any structure

[1]A case of mass abdomen diagnosed to be soft tissue sarcoma or lymphoma of the para-aortic node region by U/S proved to be a seminoma in an undescended testis. Patient said that 6 months back his right testis was removed by a 'groin' incision.
[2]Inspection and palpation of external genitalia is important in males only, **NOT TO BE DONE in females**.
[3]Don't forget the 12th man in a cricket match. He is also an important player.

KEY BOX 37.1

MOVEMENT WITH RESPIRATION

- Liver, spleen and stomach masses move freely with respiration
- Gall bladder mass also moves freely with respiration because of its proximity to liver
- Mass arising from hepatic flexure and splenic flexure of colon also have some mobility because it is in contact with liver and spleen
- Renal masses exhibit minor degree of movement with respiration because of indirect attachment to diaphragm

in contact with diaphragm moves with respiration (Key Box 37.1). For example:

- Liver, stomach, spleen, gall bladder move very well with respiration.
- Splenic flexure growth, due to contact with the lower pole of the spleen and hepatic flexure growth due to contact with liver move with respiration.
- Renal swelling moves with respiration because kidney is enclosed by fascia of Gerota which is attached above the diaphragm.

2. Finger insinuation test: This test has relevance in an upper abdominal mass.

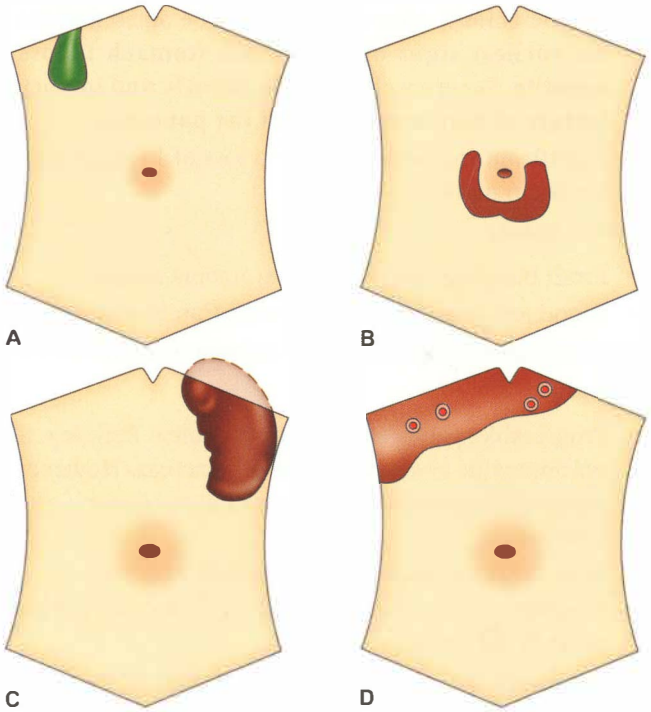
- Liver and spleen are under right and left costal margins respectively. Hence, it is not possible to get the upper margin or upper border of these organs. An attempt to invaginate between the costal margin and these masses is not possible. On the other hand, finger invagination under the costal margin is possible in a stomach mass.

3. Size, shape and surface

- An egg-shaped mass or globular mass suggests gall bladder lesion (Fig. 37.4A).
- A horseshoe shape may indicate a horseshoe kidney (Fig. 37.4B) with pathology, e.g. hydronephrosis.
- Reniform-shape suggests a renal swelling (Fig. 37.4C).
- **Large nodular surface** is seen in following conditions (Fig. 37.4D):
  - Polycystic kidney
  - Secondaries in the liver
  - Group of lymph nodes
- **Smooth surface** usually indicates a benign lesion.
  - Splenomegaly, hydronephrosis, ovarian cyst, gall bladder swelling.
- **Irregular surface** is an important feature of malignancy such as carcinoma of the stomach, carcinoma caecum.

4. Consistency

- **Hardness** is a feature of malignant lump. Thus, hepatoma, carcinoma stomach, pancreatic carcinoma present as hard



Figs 37.4A to D: Shapes of the intra-abdominal mass

lump. However, it should be remembered that often the malignant lump is firm and not hard.

- **Firm** consistency is found in ileocaecal tuberculosis, nodes of lymphoma.
- A peculiar **doughy** feel is described for tuberculous abdomen.
- It is difficult to elicit fluctuation test for intra-abdominal swellings, and often tensely cystic swellings feel firm on palpation, e.g. pseudocyst of pancreas, hydronephrosis, etc.
- Indentation or pitting on pressure can be found in a colon loaded with faeces.
- Temporary contraction of a stomach (visible gastric peristalsis) should not be confused as a **mass**.

5. Margins or borders (Fig. 37.5)

- **Upper border** cannot be made out in liver, splenic and renal swellings.
- **Lower border** is not appreciated in pelvic masses, e.g. uterine fibroid, ovarian cyst (pelvic).
- A **characteristic notch** is felt in the anterior border of splenic swelling.
- Lower border is **sharp** as in a malignant liver swelling.

6. Intrinsic mobility test (Key Box 37.2)

- An intra-abdominal mass can be mobile if it has loose attachments or if it is not within the bony cage. Thus, liver, spleen, uterine mass are not mobile because of their location within bony cage.

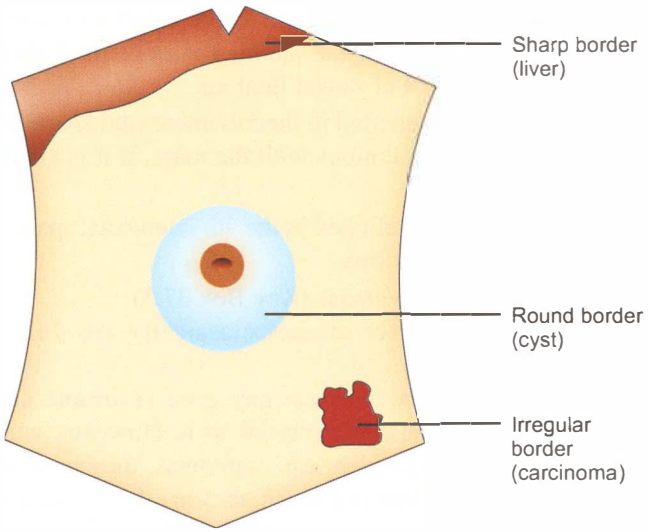
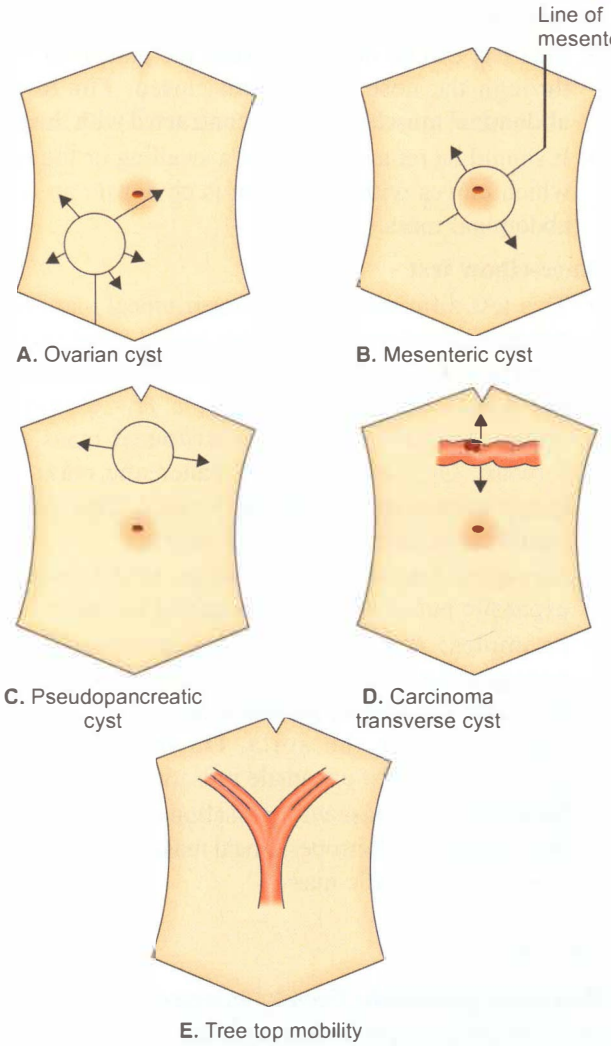


Fig. 37.5: Various types of borders



Figs 37.6A to E: Intrinsic mobility

PEARLS OF WISDOM

Check for intrinsic mobility in different positions.

- Carcinoma pyloric antrum can exhibit movements in different positions—left lateral, right lateral or even in the sitting position.
- Pancreatic carcinoma, advanced malignancies and lymph nodal masses also may not have intrinsic mobility.
- However, there are a few swellings which have characteristic mobility.

Examples

A. Ovarian cyst is a freely mobile swelling which can be moved in all directions (Fig. 37.6A).

B. Mesenteric cyst moves at right angles to the direction of the line of mesentery (Fig. 37.6B).

C. Pseudopancreatic cyst may have a minimal side-to-side mobility (Fig. 37.6C).

D. Carcinoma transverse colon has vertical mobility unless it is advanced (Fig. 37.6D).

KEY BOX 37.2

INTRINSIC MOBILITY— MASS

• Side to side	—	Gall bladder
• Vertical	—	Transverse colon
• All directions	—	Ovarian cyst
• Right angle to the direction of mesentery	—	Mesenteric cyst
• Push back to renal	—	Kidney pouch
• Tree top mobility	—	Pancreatic cystadenoma

E. Pancreatic masses: Even though they do not exhibit mobility, a cystadenoma of the pancreas because of the size and a narrow base, will exhibit **tree top mobility** (Fig. 37.6E). Any big mass abutting the undersurface of the diaphragm also moves with respiration.

F. Renal mass comes down during inspiration. As it comes down, it can be held back and can be pushed back to the renal pouch.

7. Plane of the swelling

A. Leg raising test or head raising test

- The purpose of this test is to contract rectus abdominis muscles (also other abdominal wall muscles). Intra-abdominal swellings become less prominent. On the other hand, abdominal wall swellings become more prominent, e.g. fibroma, neurofibroma, or lipoma in the abdominal wall.
- This test is done by asking the patient to raise his legs without bending at the knee (extended legs) or by raising the shoulders from the bed with arm folded over the chest.

B. Nose blowing test or straining test

- This test can be done by asking the patient to blow through the nose with mouth closed. **The lateral abdominal muscles are more contracted with this test.**
- It should be remembered that a swelling or the mass which moves with respiration is obviously an intra-abdominal mass.

C. Knee-elbow test

- This test differentiates an intraperitoneal mass from retroperitoneal mass. It is more useful when there is a **mass in the centre of the abdomen—more so in the upper abdomen**. To give a few examples, intraperitoneal cyst or intraperitoneal mass falls forward. On the other hand, pancreatic mass or a lymph node mass will not fall forward. **The test has significance only in 'selected' cases.**
- However, knee-elbow test helps to differentiate expansile pulsation from transmitted pulsation.
- Examples: A pseudocyst of pancreas will give transmitted pulsations because it overlies aorta. In the knee elbow position, pulsation disappears as it gets separated from the aorta. On the other hand, aneurysms exhibit expansile pulsations.
- Intraperitoneal mass with pulsations over it is likely to be hepatoma. Retroperitoneal mass with pulsations over it is pancreatic mass.

Special tests

1. **Bimanual palpation:** Grossly enlarged swellings may be bimanually palpable such as liver, spleen, kidney.
2. **Ballottability:** 'Ballot' means to toss about. To ballot, the swelling should be bimanually palpable and there should be a gap or space between hands which are kept anterior and posterior to the mass. Typically, renal swellings are **ballottable**. This test is done when the patient is in supine position, by keeping one hand anteriorly in the lumbar region over the swelling and the other hand posteriorly in the renal angle. A gentle push is given from behind and the swelling touches the hand which is placed anteriorly and it goes back. **Ballottability** is **because of perirenal pad of fat and due to 'pedicle'.**

Percussion

1. To demonstrate mild ascites, the patient is put in a knee-elbow position and percussion is done around umbilicus. It gives a dull note if minimal fluid is present (normally area around the umbilicus is resonant).
   - Significant or moderate fluid in the abdomen is demonstrated by percussion of the centre and flanks of the abdomen in the lying down position and in the left or right lateral position.
   - In the supine position, flanks give a dull note due to fluid. However, in the lateral position, fluid shifts down and coils of bowel float up.
2. Liver dullness is elicited in the 5th intercostal space and the dullness is continuous with the mass, if it is arising from the liver.
3. Splenic dullness is elicited in the 9th intercostal space in the left midaxillary line.
4. **Percussion** over the mass (Key Box 37.3):
   - Splenic and liver masses classically are **dull** to percuss.
   - Retroperitoneal masses may give **resonant** note because of intestines anterior to it. However, when they attain large size, e.g. sarcomas, they push the bowel to one side and hence, they are dull to percuss.
   - Stomach mass may give **impaired resonant** note because of solid growth and due to the presence of air in the stomach.
   - **Renal angle percussion:** In cases of enlargement of kidney, there will be a band of resonance anteriorly due to the colon but posteriorly it gives a dull note.
   - **Hydatid thrill:** It is demonstrated by placing 3 fingers over the swelling and percussing the middle finger. Due to the fluid in the cyst, the fluid thrill (after-thrill) is felt by the other two fingers. This clinical sign is rarely demonstrable.

KEY BOX 37.3

PERCUSSION

• Dull note	:	Liver, spleen, renal angle
• Resonant	:	Bowel anterior to the mass (e.g. retroperitoneal mass)
• Impaired	:	Stomach mass
• Shifting dullness	:	Ascites

Auscultation (Fig. 37.7)

1. Loud noisy sounds **(borborygmi)** with or without peristalsis may indicate subacute obstruction. Such patients may be having ileocaecal tuberculosis or carcinoma caecum. This should be done at right iliac fossa to listen bowel sounds.
2. Auscultation over the liver mass may reveal a **bruit** as in a rapidly growing hepatoma.
3. **Succussion splash** is a splashing sound in cases of pyloric obstruction either due to carcinoma or due to chronic duodenal ulcer.
4. Perisplenitis and perihepatitis give rise to **friction rub** as in sickle cell anaemia due to repeated infarction and adhesions.
5. Aortic aneurysm will give a **continuous murmur** in the upper abdomen.
6. **Auscultopercussion** or auscultoscraping test is done to assess lower border of the stomach or greater curvature of the stomach.

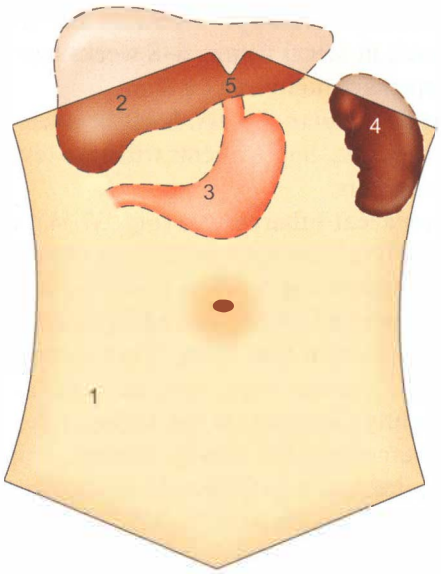


Fig. 37.7: Auscultation sites: 1–5 (*see* text in previous page)

Rectal examination

Should be done in a case of intra-abdominal mass.

1. It can detect a carcinoma or a growth in the rectum in a case of secondaries in the liver.
2. It can detect secondaries in the rectovesical pouch. **Blumer's shelf**—as hard, nodular mass and rectal mucosa is free during digital examination.

Vaginal examination

Should be done to rule out carcinoma cervix or to detect lymph nodes in the pouch of Douglas.

Bimanual examination

This should be done in cases of pelvic masses. One hand (left) is placed over the mass in the hypogastrium and right index finger or fingers inserted in the vagina or rectum in virgin females and the left hand is pressed downwards and backwards above the pubic symphysis. By this manoeuvre, details of the pelvic mass, solid or cystic, uterine or ovarian, free or fixed can be made out.

Examination of lymph nodes

In cases of abdominal masses arising from lymph nodes, a thorough search of the body should be done to rule out other group of lymph nodes such as axillary, iliac, inguinal, neck nodes (lymphoma).

Significance

- **Left supraclavicular nodes** (Virchow's) are enlarged very often in visceral malignancies mainly from gastrointestinal tract. It indicates "inoperable" nature of the disease. Entire gastrointestinal lymph drains into the thoracic duct which joins the point of confluence of internal jugular vein and subclavian vein on the left side This explains the significance of enlargement o Virchow's node. In 20% of cases, thoracic duct is singl and 10–15% of cases, it is double.
- **Significance of right supraclavicular node:** Th lymphatics from the right mediastinal lymph trunk, an from the posterior right thoracic wall which form th right upper lymph trunk drain into the commencemen of the right brachiocephalic vein.

SYSTEMIC EXAMINATION

Systemic examination should include respiratory system an cardiovascular system. Evidence of tuberculosis of the ches gives a clue about the mass in the abdomen, which may be a tubercular mass.

Differential diagnosis: Students are requested to refer clinica books for details. However, mass arising from five differen quadrants are discussed below.

MASS IN THE RIGHT ILIAC FOSSA

Parietal swelling

A. Parietal wall abscess
B. Desmoid tumour

Intra-abdominal

A. Arising from normal structures
B. Arising from abnormal structures

FROM NORMAL STRUCTURES

I. Intestines
1. Appendicular mass
2. Appendicular abscess
3. Ileocaecal tuberculosis
4. Carcinoma caecum
5. Amoeboma
6. Intussusception
7. Actinomycosis

II. Lymph nodes
1. Acute lymphadenitis
2. Lymphoma
3. Secondaries

III. Retroperitoneal structures
1. Sarcoma
2. Aneurysm
3. Iliopsoas abscess
4. Chondrosarcoma

IV. In females
1. Ovarian cyst
2. Fibroid
3. Tubo-ovarian mass

FROM ABNORMAL STRUCTURES

1. Undescended testis: Seminoma
2. Unascended kidney

Differential diagnosis of mass in the right iliac fossa (Figs 37.8 and 37.9)

I. Parietal swelling: They are extra-abdominal. On head or leg raising test, they become more prominent. They are uncommon swellings.

A. Parietal wall abscess: It is a pyogenic abscess which can occur in a haematoma, or a pyaemic abscess which can occur as a part of pyaemia as in diabetic patients. Such abscesses are very tender, with warm surface and are associated with fever, chills and rigors.

B. Desmoid tumour: It is an unencapsulated fibroma occurring in the abdominal wall.

- Occurs in multiparous females. Repeated stretching of abdominal layers (due to pregnancy) is supposed to initiate formation of tumour.
- It can also occur following abdominal wall injury including laparotomy.
- It is a firm to hard swelling.
- It has no capsule. Hence, it should be treated with wide excision.
- It does not undergo sarcomatous change.
- After wide excision, the abdominal wall has to be reconstructed by using mesh.

II. Intra-abdominal swelling:

A. Arising from structures normally present in the right iliac fossa

1. **Appendicular mass** (Fig. 37.10): It is a tender, soft to firm mass which develops 48–72 hours following acute appendicitis. It is nature's attempt to limit the spread of infection by forming a mass consisting ***of omentum, terminal ileum, caecum with pericaecal fat and inflammatory oedema.*** It is managed conservatively by Oschner-Sherren's regime because an attempt to remove the appendix may result in faecal fistula. 6–8 weeks later, an elective appendicectomy can be done.
2. **Appendicular abscess:** It will be a very tender, firm, fixed mass. Such patients will have fever with chills and rigors.
3. **Ileocaecal tuberculosis** (Fig. 37.11): Hyperplastic variety of tuberculosis forms a chronic cicatrising granulomatous reaction involving terminal ileum, caecum and part of ascending colon resulting in a mass in right iliac fossa. It is a chronic, nontender, firm, nodular mass, may have mobility, situated slightly (lumbar) on the higher side. Features of tuberculosis are usually present. It is treated by limited resection followed by ileocolic anastomosis.
4. **Carcinoma caecum** (Figs 37.12 and 37.13):
   - More common in females, around 40–50 years of age.
   - It produces bleeding per rectum, severe anaemia, etc.
   - Hard, irregular mass in right iliac fossa with fixity or restricted mobility is a usual feature. Psoas spasm indicates infiltration into psoas muscle. It is treated by right radical hemicolectomy.
5. **Amoeboma:** Can be acute or chronic. It follows an attack of amoebic typhlitis (inflammation of the caecum). Amoeboma is tender and soft to firm. It is not common to find amoebomas nowadays because of effective treatment of amoebiasis with metronidazole, tinidazole, etc.
6. **Intussusception:** Acute or chronic intussusception can give rise to a mass in the right iliac fossa which is tender and soft to firm. When acute intussusception occurs in children, it is described as idiopathic intussusception. Chronic intussusception may disappear spontaneously.
7. **Actinomycosis:** This is a rare mass in the right iliac fossa which usually develops 2–3 months after appendicectomy. A woody hard, indurated tender mass with multiple sinuses is characteristic of this condition. Sinuses discharge sulphur granules which can trickle down. Unlike tuberculosis, narrowing of lumen of the gut and lymph node enlargement does not occur.

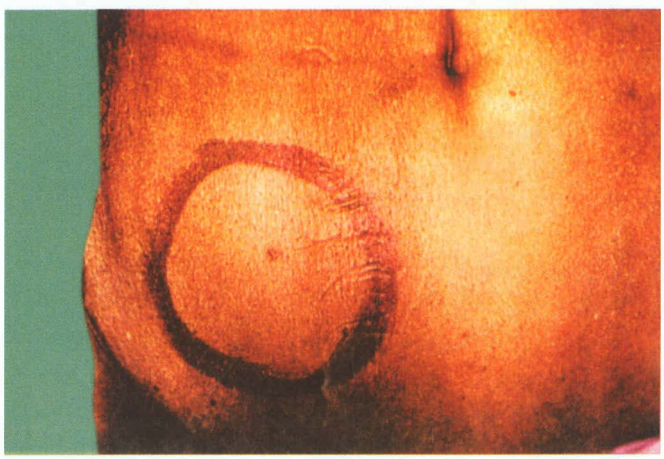


Fig. 37.8: Mass in the right iliac fossa

Fig. 37.9: Carcinoma caecum (right iliac fossa mass)

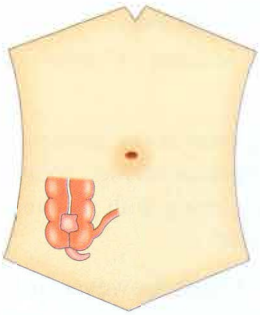


Fig. 37.10: Appendicular mass—tender, laterally placed fixed mass

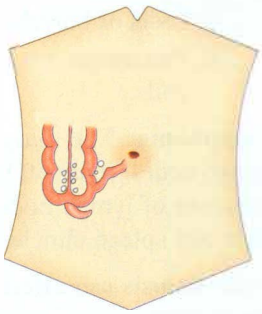


Fig. 37.11: Ileocaecal tuberculosis—caecum is pulled up and more medial

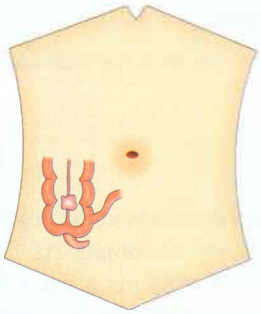


Fig. 37.12: Carcinoma caecum—irregular, nontender hard mass—more lateral

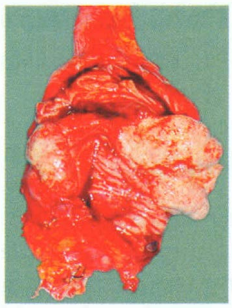


Fig. 37.13: Proliferative lesions of carcinomas produce mass

8. Lymph node mass

- Acute mesenteric lymphadenitis is common in children. It produces tender, nodular and firm mass in right iliac fossa. The child usually has fever. Acute lymphadenitis can also involve external iliac nodes as in filariasis.
- Lymphoma involving external iliac nodes, nodular, firm to hard mass with involvement of other nodes, liver, spleen, etc.
- Secondaries in lymph nodes (external iliac) from carcinoma ovary, cervix, etc. Nodes are hard and fixity is a feature.

9. Retroperitoneal sarcoma (Fig. 37.14)

- Common in young patients
- Huge, nodular, fixed lump involving lumbar, umbilical and right iliac fossa. Recent increase in size draws the attention of the patient.
- Fixed to posterior abdominal wall
- Later, obstruction of inferior vena cava results in oedema of legs.
- Pressure on the ureter can give rise to hydronephrosis.
- Liposarcoma is the commonest and may arise from pre-existing lipoma.
- Fibrosarcoma, haemangiosarcoma, leiomyosarcoma are other sarcomas.
- It is treated by wide excision followed by radiotherapy.
- Chemotherapy is also helpful, when it is not possible tc remove the entire mass.
- Debulking even if it is an advanced case is recommended.

10. Aneurysm: Iliac artery aneurysm is rare and occurs in old-aged patients. It produces a soft, pulsatile swelling in the right iliac fossa. Bruit or thrill is usually present.

11. Iliopsoas abscess (Fig 37.15)

- It is the result of tuberculosis of thoracolumbar spine. It should be suspected when a young patient complains of pain in the back referred to abdominal wall.
- Spine movements are limited.
- Gibbus is present.
- Initially, it forms paravertebral abscess and later it gravitates down beneath the medial arcuate ligament and forms psoas abscess. Psoas abscess burrows into the thigh under inguinal ligament and forms iliopsoas abscess.
- Fluctuation is present on both sides of the inguinal ligament. It is described as cross-fluctuation test (Fig. 37.16).

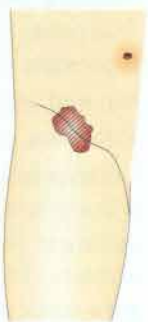


Fig. 37.15: Iliopsoas abscess

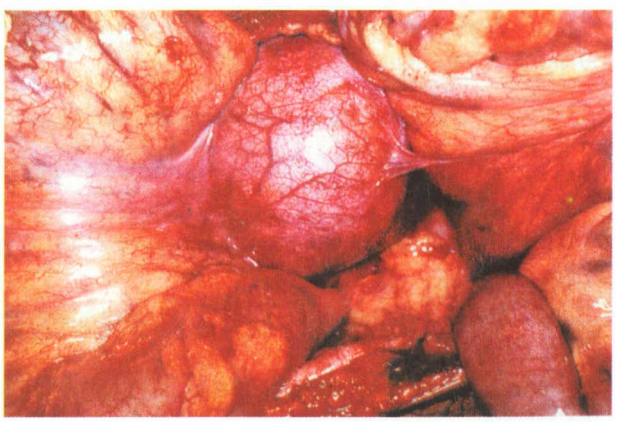


Fig. 37.14: Retroperitoneal sarcoma

Fig. 37.16: Cold abscess—the swelling was partly above and partly below the inguinal ligament

12. Chondrosarcoma of the iliac crest:

- It is a hard, fixed tumour which cannot be separated from the bone.

In females

13. Ovarian cyst: To start with, the cyst develops in the pelvis and gives rise to discomfort in the lower abdomen. As the cyst grows, it comes out of the pelvis and forms a mass in right iliac fossa. It has smooth surface, round borders, is cystic, ***freely mobile*** and can be pushed back into the pelvis. Sometimes, the cyst can attain huge size. Such freely mobile ovarian cysts have a long pedicle. Per vaginal examination gives the clue to the diagnosis.

14. Fibroid of the uterus: It presents as a firm to hard nodular mass in the suprapubic region and in the right iliac fossa.

15. Tubo-ovarian mass

- It is usually tender
- Pelvic infection is present
- It is soft to firm
- It can be bilateral

B. Arising from structures which are *not normally present*

1. Unascended kidney: It can be either in the pelvis or in the iliac fossa. Such kidney is usually not very well-developed. It presents as a lobular mass.

2. Normal mobile kidney: It can be felt in lumbar region, iliac fossa and can be pushed back into the loin.

3. Undescended testis: It is palpable in right iliac fossa *only* when it is involved by Seminoma. It is an intra-abdominal testis and is hard, irregular, fixed mass. *Absent testis* in the scrotum clinches the diagnosis. Patient may have palpable para-aortic nodes, supraclavicular nodes, etc.

FIRM TO HARD NODULAR MASS IN THE UMBILICAL REGION

1. Mass arising from lymph nodes

a. Metastasis or secondaries is one of the common lymph node masses in the abdomen. Mass can be due to para-aortic nodes from testicular tumour, melanoma, carcinoma of ovary, carcinoma of penis, carcinoma of rectum, colon, stomach in late cases.

- A para-aortic lymph node mass has the following features:
  1. Fixed
  2. Does not move with respiration
  3. No intrinsic mobility
  4. Does not fall forward
  5. Coils of bowel can be felt over the mass
  6. Percussion may be resonant because of intestinal coils.

b. Lymphoma: The mass is enlarged para-aortic group of lymph nodes. It has all features of nodes mentioned above. Presence of lymph nodes in the neck along with palpable liver and spleen clinches the diagnosis.

c. Tuberculosis can affect para-aortic nodes. However, it is uncommon.

2. Retroperitoneal sarcoma

- Common in young patients
- Rapidly growing, enlarging mass in the abdomen of short duration.
- It is firm, hard, nodular, fixed, does not fall forward and has intestinal coils anterior to it.
- Large sarcomas can cause compression on inferior vena cava or on the ureter. Therefore, pedal oedema and hydronephrosis can occur.
- Liposarcoma and fibrosarcoma are common.
- Radical surgery should be attempted and it is the only hope of cure. (Many cases may require debulking and follow-up with radiotherapy and chemotherapy.)

3. Carcinoma body of pancreas (Fig. 37.17)

- Cystadenocarcinoma of pancreas can attain a huge size. Otherwise, it is uncommon to get a large nodular pancreatic mass. However, carcinoma pancreas presenting as a palpable nodular mass indicates nonresectability. Presence of **pulsations over the mass** (transmitted) clinches the diagnosis.
- Men in the 6th decade are usually affected.
- Severe backache, loss of weight, recent development of diabetes suggest pancreatic pathology.
- Jaundice does not occur unless and until liver secondaries develop.
- It has all the features of retroperitoneal mass.
- These cases are advanced with ascites, rectovesical deposits, etc.

4. Carcinoma transverse colon (Fig. 37.18)

- Elderly patients present with constipation and bleeding per rectum.
- Firm to hard nodular mass occupying umbilical region may be found.
- It may have vertical mobility and being intra-abdominal, it falls forward.
- Caecum may be distended. Right to left peristalsis may be visible.

Mass in the umbilical region

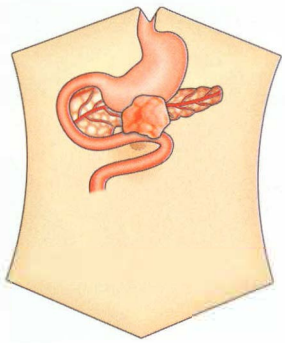


Fig. 37.17: Carcinoma body of pancreas

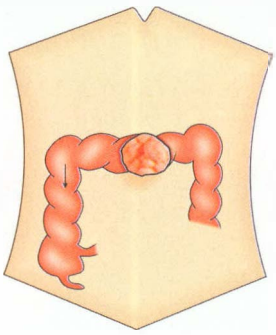


Fig. 37.18: Carcinoma transverse colon

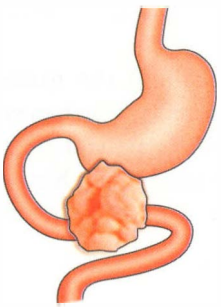


Fig. 37.19: Omental mass

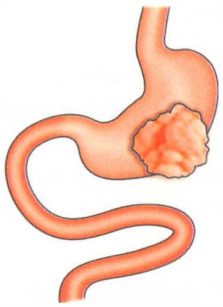


Fig. 37.20: Carcinoma stomach

Fig. 37.21: Pseudocyst pancreas

5. Tuberculous abdomen (Figs 37.19 and 37.20)

- The mass can be rolled up omentum, with lymph nodes and coils of intestines which are matted.
- This is common in children and also occur in young adults in India.
- History of evening rise in temperature, loss of weight, loss of appetite, emaciation and improper digestion gives the clue to the diagnosis.
- Ascites is present in almost all cases.
- Features of subacute intestinal obstruction can also be present.

THE CYSTIC MASS IN THE ABDOMEN

Intra-abdominal cystic swellings are interesting swellings. They occur in young children, adults, middle-aged persons. There are many cases of cystic swellings which have given a surprise at laparotomy (notoriously so in females). In children cysts have confused many competent paediatricians!! Being intra-abdominal cysts, it is not possible to elicit fluctuation and very often they are firm due to increased tension. The details of important cystic swellings are given below.

1. Pseudocyst of pancreas (Figs 37.21 and 37.22)

Tensely cystic upper abdominal mass may feel firm and tender, and does not move with respiration (*see* page 607 for details). Getting above the swelling is possible. Transmitted pulsations of the aorta can be felt over the mass which disappears on knee-elbow position. History of acute pancreatitis or blunt injury abdomen gives the clue to the diagnosis (Fig. 37.22).

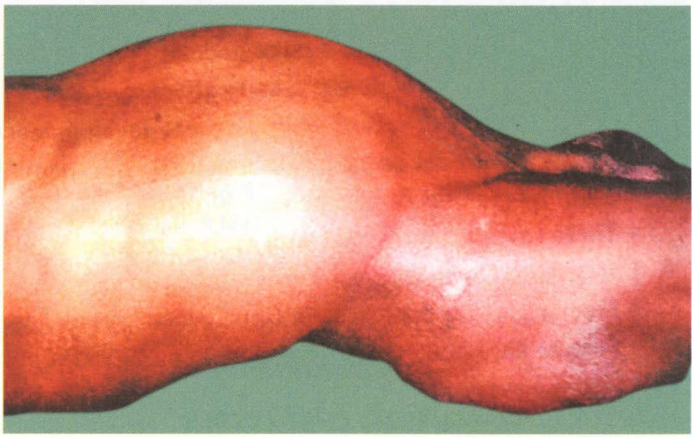


Fig. 37.22: Pseudocyst of pancreas (large)

2. Hydatid cyst of liver (Fig. 37.23)

This swelling is of long duration, is symptomless or with dull pain in the upper abdomen. The cyst is spherical with smooth surface, rounded borders and feels firm. Since it is a mass arising from liver, it moves with respiration and getting above the swelling is not possible. Classical hydatid thrill, mentioned in the books, is rarely appreciated. Simple cyst of the liver can also present as a cystic mass (Fig. 37.24).

3. Mesenteric cyst

These are congenital cysts, enterogenous or chylolymphatic, manifests in young children or during adolescence. Typically, the cyst is located in the umbilical region which moves at right angles to the direction of mesentery.

Types of mesenteric cyst (Key Box 37.4 and Figs 37.25 to 37.27, *see* page 659 also)

A. Chylolymphatic cyst is a lymphatic cyst arising from mesentery of ileum. It is a thin-walled cyst with clear fluid or chyle. It has a separate blood supply. Hence, enucleation is the treatment without sacrificing the bowel.

B. Enterogenous cyst is a duplication cyst from the intestine or due to diverticulum of the mesenteric border of the intestine. It is thick walled and contains mucus. This cyst is treated by excision of cyst with bowel segment because both share the same blood supply.

Complications

- Torsion of the cyst resulting in acute abdominal pain.
- Rupture of the cyst due to trauma.
- Haemorrhage into the cyst.

Fig. 37.23: Hydatid cyst

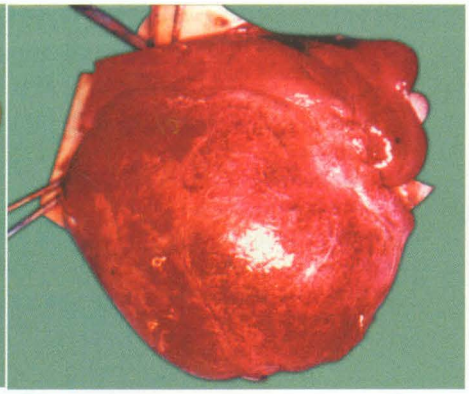


Fig. 37.24: Intra-abdominal cyst: Serous cyst in the liver

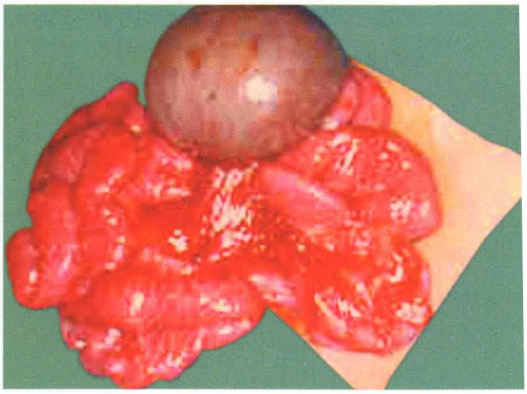


Fig. 37.25: Enterogenous cyst at surgery

KEY BOX 37.4

MESENTERIC CYST—TYPES

- Chylolymphatic cyst
- Enterogenous cyst
- Urogenital remnant
- Teratomatous dermoid cyst

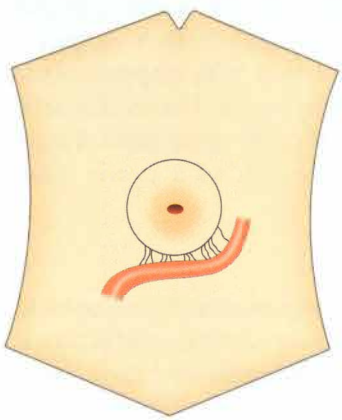


Fig. 37.26: Chylolymphatic cyst—it can be excised without resection of the bowel

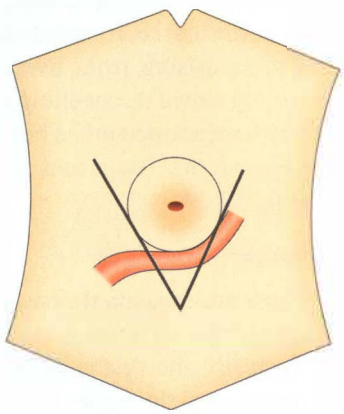


Fig. 37.27: Enterogenous cyst—it requires excision of intestine also along with the cyst

4. Hydronephrosis

Large hydronephrosis can attain a huge size without producing any symptoms. Bulk of the swelling is confined to one side of abdomen, with prominent bulge in the loin. It is difficult to elicit fluctuation in a tensely cystic intra-abdominal mass. Bimanual palpation and ballotability give the clue to the diagnosis. One of the large cysts of polycystic kidney can present as a large renal cyst.

5. Ovarian cyst

It is a freely mobile, firm or soft mass in any quadrant of the abdomen. Such ovarian cysts, once they come out of pelvis will have free mobility. On pushing **the mass upwards there will be traction** on the pedicle, which may result in pain (Fig. 37.28). In any female patient who presents with lower abdominal mass, ovarian mass has to be considered first, and only then consider other possibilities.

6. Retroperitoneal lymphatic cyst

Retroperitoneal cyst is one of the commonest of lymphatic cysts, which grows slowly to attain large size. Typically it is painless, seen in young patients and is tensely cystic. The bowel loop may be felt over the mass (retroperitoneal mass), or bowel loops may be pushed to the side.

7. Encysted ascites

This consists of ascitic fluid loculated by many loops of intestine along with omentum. Loss of weight, fever, anorexia, emaciation are the other features.

8. Abdominal aortic aneurysm (AAA)

- Majority of cases are due to atherosclerosis and most of the aortic aneurysms are infrarenal. Hence, they present

Fig. 37.28: Ovarian cyst

with a swelling in the umbilical region or epigastric region and are associated with backache (Key Box 37.5).

- Often they contain clotted blood. Hence, they feel firm, not compressible, fixed and tender.
- Characteristic feature of an abdominal aortic aneurysm is **expansile pulsation**. This can be appreciated by palpating the swelling gently all around. In the knee-elbow position, the pulsations do not disappear. (Transmitted pulsations disappear in the knee-elbow position.)
- The **femoral pulses** may be normal unless there is thrombosis or rupture of the aneurysm, giving rise to features of acute ischaemia.
- Pressure effects such as **venous oedema** due to pressure on the inferior vena cava or erosion of vertebrae may be found.
- Ultrasound to confirm the aneurysm and also to rule out suprarenal aneurysm.
- It is treated by repair of aneurysm, by incising the aneurysm and suturing a dacron graft end to end, inside the aneurysmal sac.

9. Rare cystic swellings in the abdomen (Fig. 37.29)

- **Omental cyst:** This is usually a lymphatic cyst which occurs in children and can attain a huge size. Sudden enlargement indicates haemorrhage. Excision is easy.
- **Large mucocoele** of the gall bladder can present as a tense cystic, slightly tender mass in the upper abdomen.

KEY BOX 37.5

AORTIC ANEURYSM

- Elderly males > 60
- Hypertensive
- Expansile pulsation +
- Anterior rupture: 20%—haemoperitoneum
- Posterior rupture: 80%—retroperitoneal haematoma
- > 6 cm size—dangerous

PEARLS OF WISDOM

One should not forget distended bladder in the lower abdomen as a cause of cystic swelling.

MASS IN THE EPIGASTRIUM

Mass in the epigastrium is one of the common long cases kept in the examination. Students should consider mass arising from liver and stomach first. Other possibilities must be considered later because common cases are common.

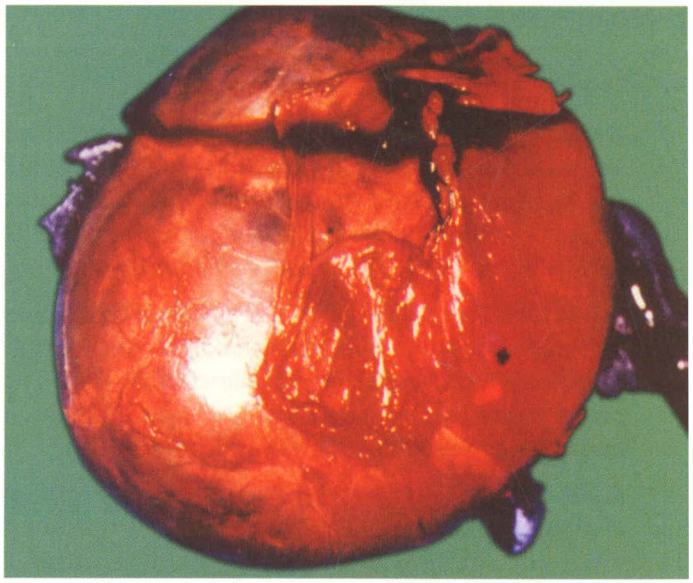


Fig. 37.29: Large lymphatic cyst arising from omentum

CLINICAL NOTES

A female child, aged 6 years, was examined by a paediatrician for generalised abdominal distension. All the investigations were normal. The child was put on antituberculous treatment, as the treating paediatrician diagnosed this case as tuberculous ascites. Child was brought back after 9 months with no improvement and having abdominal pain since 2 days due to sudden increase in the size of the swelling. A paediatric surgeon was consulted, who palpated the abdomen and said she does not have ascites but has a cyst and that the wall of the thin cyst can be felt. At exploration, a large cyst arising from the omentum and occupying all the 9 quadrants of the abdomen, was excised. Histopathological report was lymphatic cyst (Fig. 37.29).

CLASSIFICATION

I. MASS ARISING IN THE ABDOMINAL WALL

- First do the head raising test. If the mass becomes more prominent, it is extraperitoneal (abdominal wall).
- Lipoma, neurofibroma or desmoid tumour arising from the rectus sheath can present as a mass in the epigastrium.
- Also note epigastric hernia occurs in this region. It is a hernia, not a mass.

PEARLS OF WISDOM

Any hard subcutaneous swelling in the abdominal wall of recent origin can be a metastasis.

II. INTRAPERITONEAL MASS

1. Mass arising from the liver

A. Hepatoma (Fig. 37.30): Liver is enlarged, hard, irregular, and nontender. However, rapidly growing hepatomas are tender, firm and even a bruit is heard over the swelling. Rapid deterioration of health in a cirrhotic patient is usually due to the development of a hepatoma.

B. Secondaries in the liver (Fig. 37.31): Usually both lobes are enlarged, have nodular surface without a bruit. Jaundice is a late feature in secondaries of the liver. The primary may be obvious as a colonic mass, a stomach mass or a testicular tumour, etc. (*see* page 533).

C. Hydatid cyst (Fig. 37.32): It is a benign swelling. History of contact with a dog is usually present. Epigastric swelling is due to enlarged liver which is smooth or irregular, nontender with rounded borders. Classical hydatid fremitus and thrill are rarely elicited. General health of the patient is usually good.

D. Simple cyst (Fig. 37.33): It is not a clinical diagnosis but is mentioned here only for discussion. It is a serous cyst. Single big cyst can also be a part of polycystic disease of the liver.

2. Mass arising from the stomach

- For all practical purposes, the only mass arising from the stomach in the epigastrium is carcinoma stomach. It is hard, irregular and moves with respiration. Usually the patient is a male with loss of appetite and weight. Vomiting is a feature. If there is a growth in the pyloric antrum, visible gastric peristalsis can be seen in the epigastrium. (Students are hereby requested **not to offer lymphoma of the stomach or GIST** of the stomach as a **clinical diagnosis unless asked for by the examiner,** for a differential diagnosis.)

3. Omental mass

- Omentum gets involved in tuberculosis as a firm, nodular mass (Fig. 37.34) or in secondaries from intra-abdominal malignancies as a hard, nodular mass (Fig. 37.35). Classically it moves with respiration.
- Rarely, omental cyst can present as a tensely cystic mass in the epigastrium.

III. RETROPERITONEAL MASS

A. Pseudopancreatic cyst (Fig. 37.36): It forms a tense cystic mass, felt as firm mass in the epigastrium. Its upper border can be made out. It does not usually move with respiration. It has smooth surface and round borders. History of acute pancreatitis or blunt injury abdomen is usually present. Pulsations over the mass (transmitted) suggest that it is a mass close to the aorta. In such a case, it is a pseudocyst. Gurgle heard anteriorly suggests distended stomach.

B. Cystadenoma: Cystadenomas of pancreas are benign and can attain huge sizes. It can present as a mass in the epigastrium, left hypochondrium or umbilical region. They exhibit what is described as 'tree top mobility'.

C. Carcinoma body of pancreas can present as a mass in the lower part of epigastrium or upper umbilical region. The mass is hard, irregular, fixed and does not move with respiration. Presence of severe backache and loss of weight are important features.

D. Abdominal aortic aneurysm (AAA) (Fig. 37.37): An elderly patient, usually a hypertensive presents with features of abdominal pain, swelling or features of ischaemia of the lower limb. On examination, tender swelling in the epigastrium with a characteristic expansile pulsation is present. Knee-elbow test will help differentiate it from transmitted pulsations. Presence of a bruit and weak or absent lower limb pulses (due to thrombus) also helps in establishing the diagnosis.

E. Lymph node mass (Fig. 37.35)

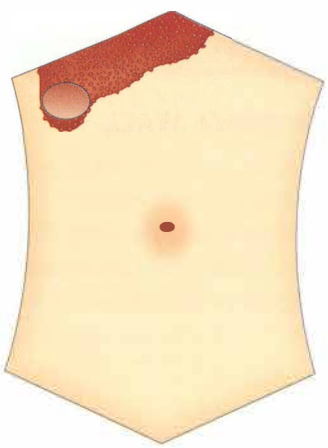


Fig. 37.30: Hepatoma

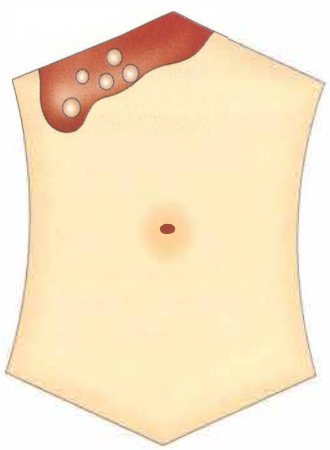


Fig. 37.31: Secondaries in the liver

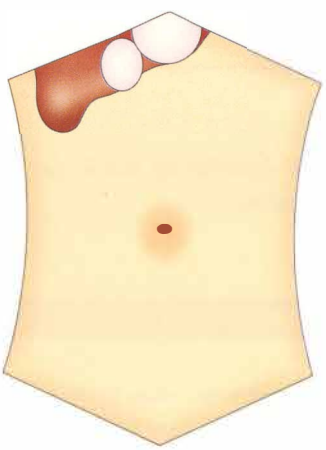


Fig. 37.32: Hydatid cyst

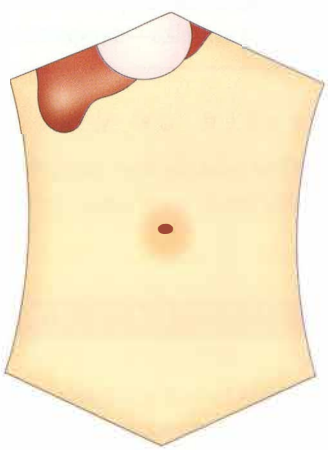


Fig. 37.33: Simple cyst

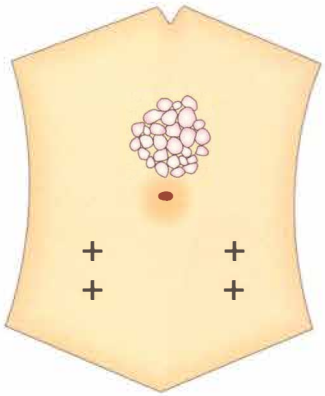


Fig. 37.34: Tuberculous abdomen—ascites and rolled up omentum

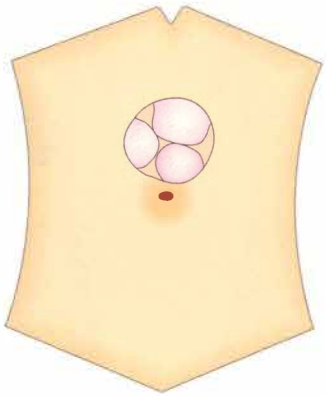


Fig. 37.35: Lymph nodes nodular, firm to hard fixed

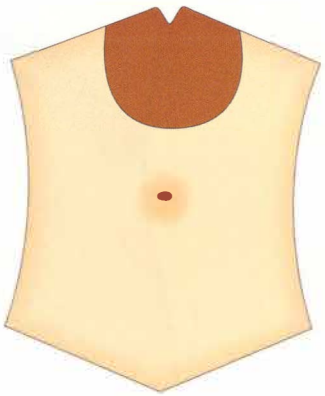


Fig. 37.36: Pseudocyst of pancreas: Large enough— in contact with diaphragm

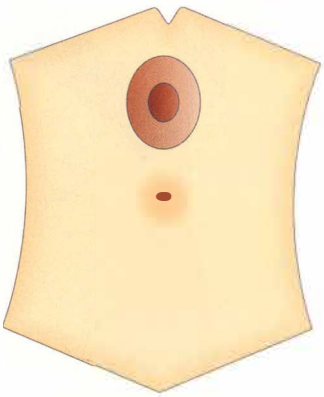


Fig. 37.37: Abdominal aortic aneurysm: Pulsatile firm mass

MASS IN THE RIGHT HYPOCHONDRIUM (Fig. 37.39)

I. Parietal: On head raising test, the lump becomes more prominent.

A. Lipoma, neurofibroma: They can be part of multiple lipomatosis or multiple neurofibromatosis. If pain and pigmentation are present, it is neurofibroma.

B. A hard nodule in the parietal wall can be due to

a. **Secondary deposit** in the skin/subcutaneous tissue specially when skin is infiltrated and ulcerated. Common primaries are malignant melanoma, bronchogenic carcinoma, hepatoma.

b. **Non-Hodgkin's lymphoma:** 'T' cell type

c. **Cold abscess:** Spine tenderness with or without history of tuberculosis gives the clue to the diagnosis.

II. Intra-abdominal swellings: On head raising test, the lump becomes less prominent.

1. Liver: Only chronic masses are discussed

A. Secondaries in liver (Fig. 37.38 and Key Box 37.6)

- Entire liver is enlarged (both lobes)
- Nodular surface
- Sharp border
- Hard in consistency
- Rare umbilication sign, evidence of primary, emaciated patient, poor health, loss of appetite and weight are other features.

B. Hepatoma (Key Box 37.7)

- One lobe is enlarged
- Firm to hard, irregular
- Very tender liver
- Bruit/thrill may be present
- Evidence of chronic liver disease such as serum hepatitis or cirrhosis is usually present.

C. Polycystic disease of the liver

- Both lobes are enlarged
- Nodular

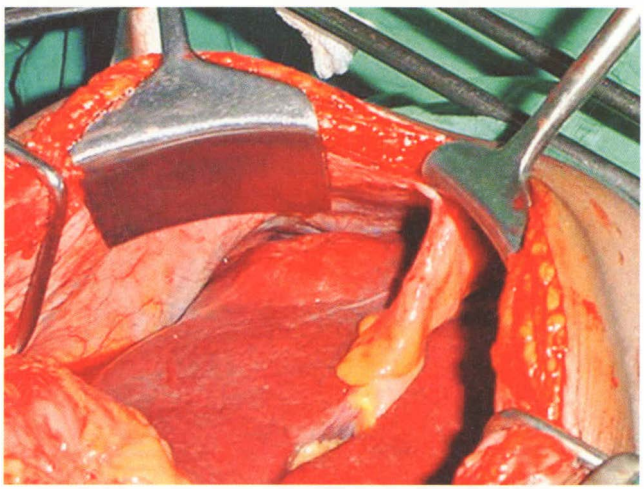


Fig. 37.38: Large secondary in the liver from carcinoma stomach

KEY BOX 37.6

ANATOMIC FEATURES OF LIVER MASS

- Location: Hypochondrium (right and left) and epigastrium
- Moves with respiration
- Finger—insinuation between costal margin and mass is not possible
- No intrinsic mobility and dullness
- Dull note over the liver which will continue with the mass

KEY BOX 37.7

TENDER LIVER MASS

- Hepatoma
- Amoebic liver abscess
- Suppurative pylephlebitis
- Congestive cardiac failure
- Infected hydatid cyst

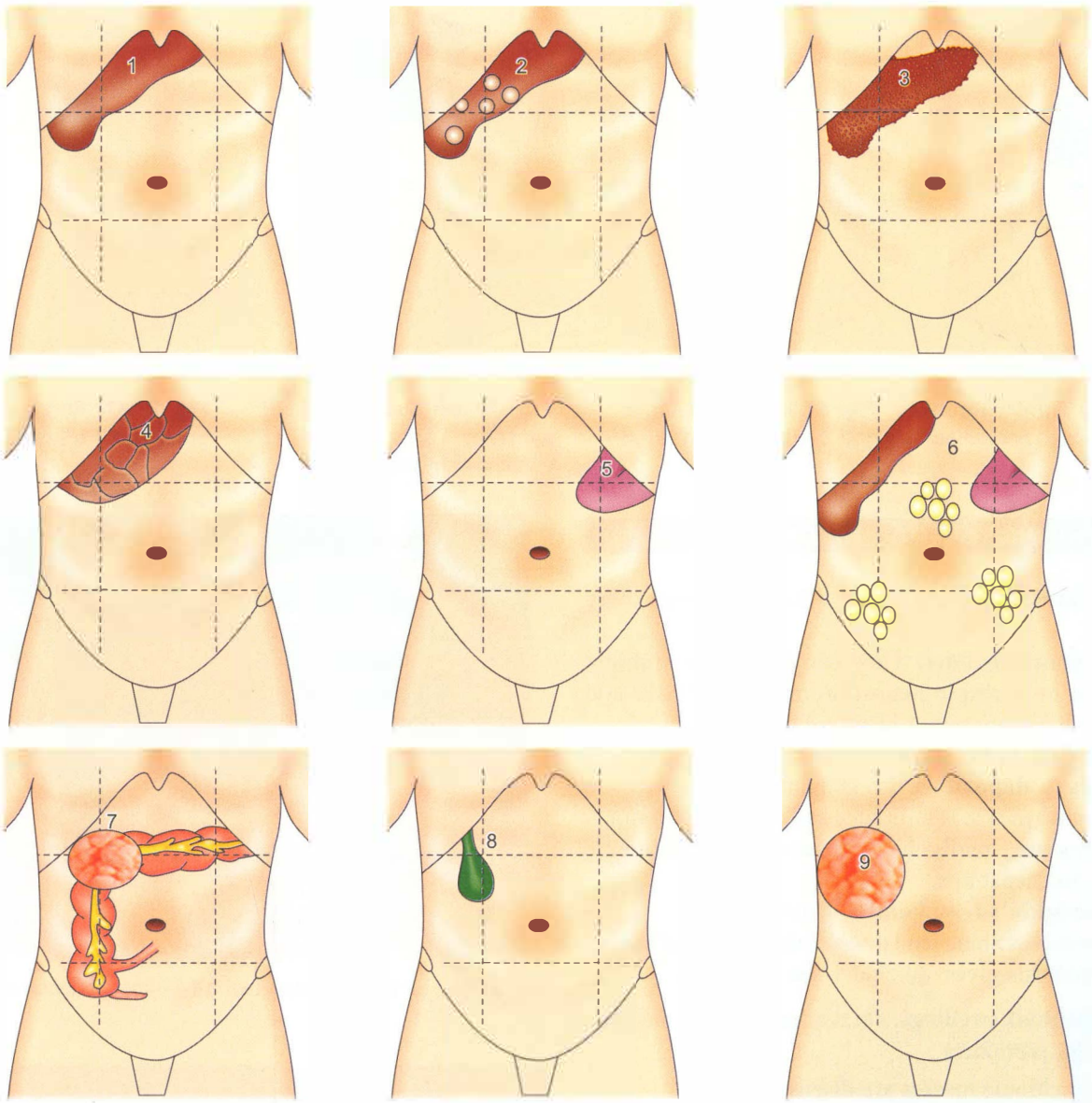


Fig. 37.39: Differential diagnosis of mass in the right hypochondrium: (1) Smooth hepatomegaly in lymphoma or due to medical causes, (2) secondaries in the liver—hard and nodular, (3) hepatoma—irregular, hard or firm, (4) polycystic disease of the liver—firm and nodular with round borders, (5) with splenomegaly—could be portal hypertension, (6) hepatomegaly, splenomegaly, para-aortic lymph nodes and iliac nodes—Hodgkin's lymphoma, (7) carcinoma ascending colon—hard irregular mass, (8) palpable gall bladder—smooth, round borders, and (9) renal mass

- Nontender
- Round borders
- General health is good

The patient would have presented to the hospital with pain due to haemorrhage in a cyst.

D. Hydatid cyst

- One or both lobes are enlarged
- Smooth or nodular surface
- Round borders, nontender
- General condition of the patient is good
- **Hydatid thrill—rare 'physical sign'** may be present.

E. Cirrhosis of liver

- Liver may be enlarged: Firm and irregular in pre-cirrhotic cases. Splenomegaly, ascites will help in the diagnosis.
- Other features of liver cell failure such as gynaecomastia, spider naevi, palmar erythema may be present.

F. Lymphoma

- Liver is palpable, one or two finger-breadths, firm or hard, smooth or irregular, nontender.
- Splenomegaly and lymphadenopathy will help in the diagnosis.

G. Congenital Riedel's lobe

- It is a tongue-shaped projection from the inferior border of liver. It is on the right side, can be mistaken for gall bladder.

2. Gall bladder mass (Key Box 37.8)

Causes of gall bladder enlargement

- **Back pressure:** Distal obstruction periampullary carcinoma. Such gall bladder is firm, smooth and associated with jaundice.
- **Carcinoma gall bladder:** Hard, irregular, fixed
- **Acute cholecystitis:** Tender, vague, well-defined mass.
- **Mucocoele:** Nontender, palpable gall bladder without jaundice
- **Empyema:** Very tender, gall bladder mass

3. Colonic mass

A. Carcinoma hepatic flexure

- Firm to hard irregular mass
- Restricted mobility
- Moves with respiration because of its contact with liver
- Resonant or impaired resonant note on percussion (liver is dull on percussion)
- Caecum may be distended if there is obstruction.

B. Large ileocaecal tuberculosis

with pulled up caecum may also be palpable. Such masses may be bimanually palpable but not ballotable.

4. Renal mass

- Importantly renal mass is palpable mainly in the **lumbar region, loin** and in the right hypochondrium.
- Carcinoma kidney is hard and irregular
- Upper border is usually not palpable—it is under cover of the 12th rib.
- Hydronephrotic kidney will be firm and smooth

5. Suprarenal mass

- Clinically they have all features of a renal mass
- Hence, symptoms of the patient may have to be correlated. A few examples are given here:
  - Cushing's syndrome
  - Phaeochromocytoma

KEY BOX 37.8

CLINICAL FEATURES OF A GALL BLADDER MASS

a. It is oval, e.g. egg-shaped swelling
b. It is tense. Hence, feels more firm in consistency
c. Moves freely up and down with respiration—better seen in thin patients
d. May have slight side-to-side mobility
e. It is felt slightly posterior to (step deep) inferior border of the liver

PEARLS OF WISDOM

Offer renal mass as diagnosis. When asked for differential diagnosis, mention suprarenal mass.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated.
- Mass in the right hypochondrium has been added.
- New simple diagrams have been added.

MULTIPLE CHOICE QUESTIONS

1. The most diagnostic sign of a renal mass is:
A. Moves with respiration
B. Upper pole cannot be felt
C. It enlarges downwards
D. Ballotability

2. Which one of the following lower horizontal lines divides abdomen into regions?
A. Transpyloric line
B. Transcolic line
C. Transtubercular line
D. Transanterior superior iliac spine line

3. Which of the following masses does not move with respiration?
A. Kidney
B. Hepatic flexure
C. Tail of the pancreas
D. Para-aortic lymph node mass

4. Notch is a diagnostic sign of which mass?
A. Spleen B. Liver
C. Kidney D. Adrenal gland

5. Which of the following does not have intrinsic mobility?
A. Fibroadenoma breast
B. Ovarian cyst
C. Mesenteric cyst
D. Multinodular goitre

6. The following are true for renal masses *except:*
A. It is bimanually palpable
B. It is ballotable
C. It does not move with respiration
D. Upper border is usually not felt

7. Blumer's shelf refers to:
A. Rectouterine pouch
B. Rectovesical pouch
C. Rectosacral pouch
D. Rectoprostatic pouch

8. Following tumours can occur in the abdominal wall *except:*
A. Desmoid tumour
B. Endometriosis
C. Dermoid tumour
D. Fibromatosis

9. Following appendicectomy, after 2 months, if wood indurated mass develops in the right iliac fossa wit sinuses, what is the diagnosis?
A. Tuberculosis
B. Crohn's disease
C. Amoeboma
D. Actinomycosis

10. Acute intussusception mass has following feature *except:*
A. Mass is tender
B. Mass is felt in the umbilical region
C. It is a sausage-shaped mass
D. In the right iliac fossa caecum gurgles

11. Which of the following masses does not have cross fluctuation?
A. Iliopsoas abscess
B. Plunging ranula
C. Collar stud abscess
D. Branchial cyst

12. Following are true for retroperitoneal sarcoma *except:*
A. Common in young patients
B. Mass does not move with respiration
C. It can attain a hard and large mass
D. Free fluid is usually present in the abdomen

13. The diagnostic feature of mesenteric cyst is:
A. It is present in the umbilical region
B. It is dull to percuss
C. It falls forward
D. Moves at right angle to the direction of mesentery

14. Which of the following masses is nontender?
A. Hepatoma
B. Appendicular mass
C. Carcinoma stomach
D. Cholecystitis

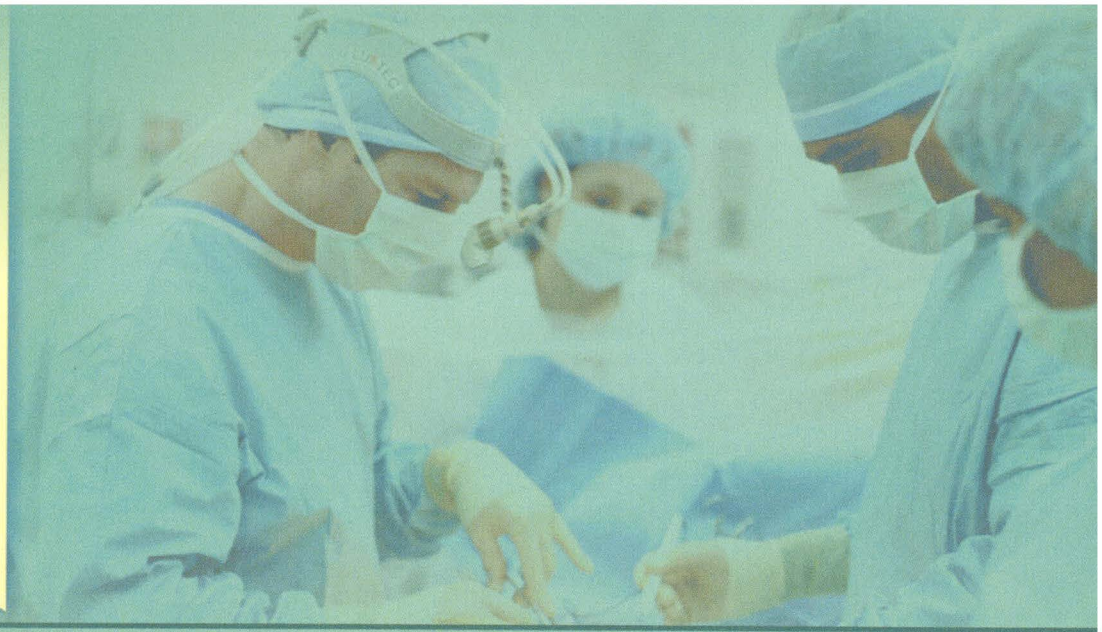
15. Murphy's triad of symptoms include the following *except:*
A. Pain
B. Vomiting
C. Fever
D. Jaundice

ANSWERS

1 D	2 C	3 D	4 A	5 D	6 C	7 B	8 C	9 B	10 A
11 D	12 D	13 D	14 C	15 D					

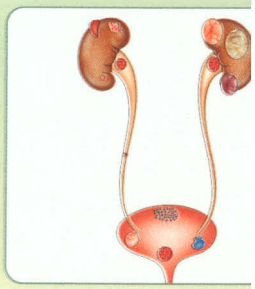
Section

III



Urology

38. Investigations of the Urinary Tract
39. Kidney and Ureter
40. The Urinary Bladder and Urethra
41. Prostate and Seminal Vesicles
42. Penis, Testis and Scrotum
43. Differential Diagnosis of Haematuria



38

Investigations of the Urinary Tract

- Urine
- Blood
- Intravenous urogram
- Retrograde pyelography
- Renal arteriography
- Cystourethrography
- Urethrography
- Ultrasonography
- Computerised tomography
- Radioisotope scanning
- Endoscopy
- What is new?/Recent advances

URINE EXAMINATION

Specific gravity: It varies from 1.005 to 1.040 according to the state of hydration of the patient. In **chronic renal failure,** the concentrating ability of the kidneys are lost and the specific gravity remains fixed at **1.010**.

pH: The urinary pH normally ranges from 4.5 to 8. It varies depending on serum pH. Urine pH influences the type of stone formed, e.g. **alkaline urine**—infection by **urea splitting organisms**, **infection stones are formed** and in **acidic urine, uric acid** and cystine stones occur.

Normally, urine is devoid of blood, protein and sugar.

Proteinuria: It is defined as **excretion of protein > 150 mg/day**. Protein, especially albumin, appears in urine following exercise, glomerulonephritis and nephrotic syndrome.

Sugar: It appears in **urine in diabetes mellitus**. Transient postprandial glycosuria may be seen when serum glucose levels exceed the renal threshold.

Blood: This is seen as presence of red blood cells (RBCs) in urine. Presence of more than 3 **RBCs/high power field is considered significant**. Haematuria, haemoglobinuria and myoglobinuria give rise to red colour in urine.

The causes of haematuria may be **glomerular or nonglomerular**. Glomerular causes are nephritis, analgesic nephropathy, IgA nephropathy and connective tissue disorders. Nonglomerular causes are stones or tumours involving the urinary tract (e.g. renal cell carcinoma, transitional cell carcinoma). More details are given in Chapter 43, page 1002.

Ketones: Are seen in urine in diabetic ketoacidosis, starvation and pregnancy.

White blood cells (WBCs): Presence of WBCs in urine is called pyuria (>5 cells/hpf). The causes are urinary tract infection, stones, glomerulonephritis, foreign bodies, tuberculosis (sterile pyuria) and malignancies (sterile pyuria).

CASTS AND CRYSTALS

Casts: Tamm-Horsfall protein is a mucoprotein from renal tubular cells. It forms nucleus of all casts. It entraps RBC/WBC/epithelial cells to form respective casts. Hyaline casts are without cells and are normal.

Crystals: Different types of crystals are formed in acidic and alkaline urine and act as precursors for stone formation. Calcium phosphate and struvite crystals form in alkaline urine. Calcium oxalate, uric acid and cystine crystals form in acidic urine.

URINE CYTOLOGY

Urine cytology refers to identification of malignant cells under the microscope. If positive, it indicates transitional cell carcinoma of bladder. Cytology is not useful in the detection of other types of carcinoma such as squamous cell or adenocarcinoma.

24-hour urinary studies

These are indicated in metabolic evaluation of stone disease (identifies various abnormalities of electrolyte homeostasis) and in the evaluation of recurrent pyelonephritis in children and diabetic nephropathy (degree of proteinuria).

BLOOD TESTS

- ***Prostatic surface antigen (PSA):*** A glycoprotein, it is elevated in prostatic diseases. Its estimation is used for screening of carcinoma prostate but is not specific for this condition. Normal levels 0–4 ng/ml. Carcinoma should be suspected if levels exceed 4 ng/ml (*see* page 977).
- ***Testicular tumour markers***
  Alphafetoprotein (AFP): Elevated in embryonal carcinoma, yolk sac tumour
  β-human chorionic gonadotropin (β-hCG): Very high levels in choriocarcinoma
  Lactate dehydrogenase (LDH): Also elevated in embryonal carcinoma and seminoma. It indicates bulk of the disease or tumour burden. It is helpful in diagnosis, staging, follow up during/after treatment and prognostication.
- ***Sex hormones*** such as follicular stimulating hormone (FSH), testosterone and prolactin may also be measured.

X-RAY KUB

- Plain X-ray KUB is a baseline investigation in suspected cases of calculous disease.
- It should be taken in supine position which should cover pubic symphysis and lower two ribs.
- A fat free, low residue diet, dimol 2 tablets, 3 times daily for 2–3 days prior to the X-ray is required.
- Stones appear as a **white** shadow (images are given in renal stones, *see* page 936, 937)

IMAGING

Today renal imaging (Fig. 38.1) is the most important part of investigating a case of urinary tract. There are many investigations, important ones being given below:

Iodinated contrasts

Ionic: Diatrizoate, metrizoate, ioxaglate (more side-effects, cheaper).
Nonionic: Iopamidol, iopromide (fewer side-effects, costly).

INTRAVENOUS PYELOGRAPHY (IVP) AND INTRAVENOUS UROGRAM (IVU) (Figs 38.1 to 38.5)

Aim

1. To study renal function
2. To detect any pathology of kidneys, ureters and bladder
3. To study any anatomical variations of the renal system.

Fig. 38.1: IVU 5 minutes picture

Procedure

- A fat-free, nonresidual diet is given for 2 to 3 days prior to the procedure to avoid intestinal gas shadows.
- Dimol 2 tablets, 3 times daily for 2–3 days prior to the procedure to expel the gas.
- The patient should not take oral fluids 6 hours before the procedure.
- Radiological contrast dye: 45% sodium diatrizoate, 20–40 ml is injected through median cubital vein.

Requirements before IVP

1. Normal renal function is a prerequisite for IVU. Serum creatinine is reliable, not urea because of variations in urea levels depending on hydration. Normal value of serum creatinine is 0.5 to 2 mg%.
2. Plain X-ray kidney ureter bladder (KUB) region to look for a renal stone—90% of the renal stones are radio-opaque (only 10% of gall stones are radiopaque).
   - To distinguish between renal stones and gall stones on plain abdominal radiograph, in case of doubt, take lateral film, stones anterior to the vertebral column are gall stones and posterior to it are renal stones.

Precautions while injecting the dye

1. The dye should be given very slowly
2. The dye should not extravasate
3. If bronchospasm occurs, in addition to inhale bronchodilator, hydrocortisone 100 mg and an antihistaminic should be administered IV.
4. In cases of urticaria and skin rashes, an antihistaminic must be given.

Radiography

1. Early films taken after 2 or 5 minutes, demonstrate the kidney outline (nephrogram).

Fig. 38.2: IVU after 20 minutes

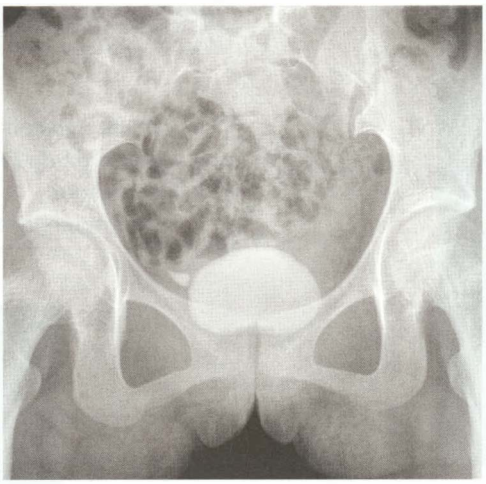


Fig. 38.3: Observe bowel gas, inadequate preparation

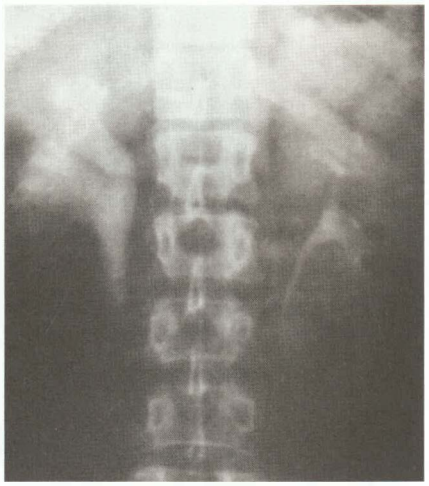


Fig. 38.4: IVU showing hydronephrosis

2. 5 minutes later, pelvicalyceal system is visualised (Fig. 38.1).
3. 15–20 minutes later, ureter, bladder can be visualised (Fig. 38.2).
4. Post-voiding picture is taken to demonstrate any residual contrast in the urinary bladder.
   - Abdominal compression has to be applied to demonstrate pyelograms better.

Contraindications for IVP

1. Idiosyncrasy to iodine: Test dose should be given before hand.
2. Renal failure: Kidneys fail to excrete the drug.
3. Multiple myeloma: The dye precipitates myeloma proteins, blocks the ureter and kidney, and causes anuria.
4. Hyperuricaemia: Uric acid crystals deposit in the renal tubules.
5. Sickle cell anaemia: Precipitates sickle cell crisis.
6. Dehydration.

Uses of IVU

1. Diagnosis of congenital abnormalities such as polycystic kidney, horseshoe kidney, single kidney, duplication of kidneys and ureters.
2. Diagnosis of hydronephrosis, hydroureter (Fig. 38.4).
3. Obstruction to pelviureteric junction, ureters, primary obstructed megaureter.
4. Diagnosis of renal, ureteric stones and bladder stones.
5. To diagnose renal tuberculosis, tumours.

Intraoperative one-shot IV pyelogram

In ureteral injuries, when delayed contrast images are not possible because of haemodynamic instability, an intra-operative one-shot (2 mg/kg IV contrast material given 10 min before flat plate abdominal X-ray) IV pyelogram (IVP) is recommended in patients with hypotension or a history of significant deceleration, despite absence of gross haematuria.

Fig. 38.5: IVU showing double ureter

RETROGRADE PYELOGRAPHY (RGP) OR RETROGRADE URETEROGRAPHY (RGU)

Indications

1. When the kidney is not visualised by IVU
   a. Gross hydronephrosis (Fig. 38.6)
   b. Very high blood urea
2. To selectively collect the urine sample from renal pelvis, e.g. renal tuberculosis.
3. History of allergy to intravenous contrast materials
4. Prior to ureteroscopy.

Procedure

- A cystoscopy is done first.
- Ureteric orifices are identified and cannulated by a flexible, catheter which is introduced up to the pelvis of kidney and

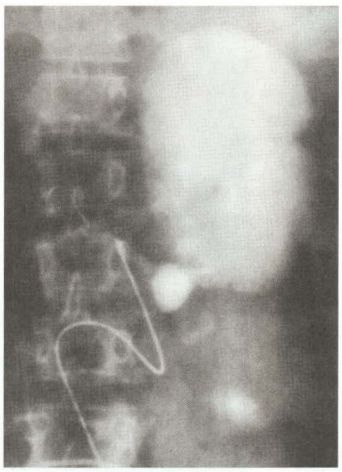


Fig. 38.6: RGP: Retrocaval ureter with hydronephrosis

the dye is injected. X-rays are taken at 5 minutes, 15 minutes and 30 minutes.

Uses

1. Anatomical evaluation of pelvicalyceal system.
2. Early diagnosis of renal tuberculosis.
3. Since the dye is injected directly into the pelvis, pelvicalyceal system can be identified better which helps in the diagnosis of **early transitional cell carcinoma of kidney.**

Complications of RGP

1. It is an invasive procedure and hence, urinary tract infection can occur. Prophylactic antibiotics are given before the procedure.
2. Rarely chances of perforation of the bladder or perforation of the ureters may occur.

RENAL ARTERIOGRAPHY: ANGIOGRAPHY

Technique

The technique used now is **digital subtraction angiography (DSA)**. There are two methods in which the test can be performed.

1. **Retrograde arteriography** using Seldinger technique. Selective renal angiography can be done by using a catheter over a guide wire passed into renal artery.
2. **Translumbar aortography** is done wherein aorta is punctured with a needle from behind, above the renal arteries at the level of 1st lumbar vertebra.

Dose

For aortography 30 ml of contrast (hypaque) and for selective renal angiography 6–8 ml is used.

Uses (Key Box 38.1)

1. To demonstrate pathological anatomy of the renal artery when renal artery stenosis or aneurysm is suspected.

KEY BOX 38.1

COMPLICATIONS

1. Tubular necrosis of the kidney
2. Paraplegia due to spasm of spinal arteries
3. Haematoma
4. Thromboembolism

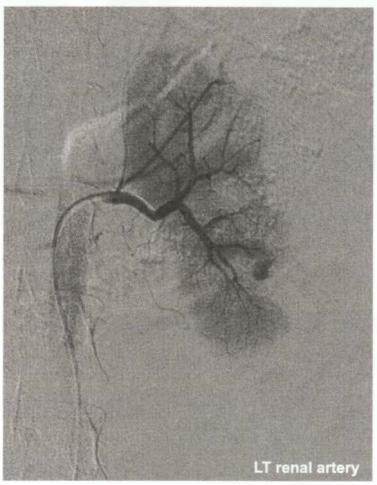


Fig. 38.7: Renal angiogram—aneurysm

2. In renal cell carcinoma, tumour vascularity and extension of the tumour into the renal vein can be diagnosed during the venous phase.
3. Bleeding from the kidney due to trauma, post-percutaneous nephrolithotomy (PCNL) bleeding or arteriovenous malformation.
4. Therapeutic application:
   - Transluminal angioplasty can be done by inflating the balloon in cases of renal artery stenosis.
   - Embolisation of bleeding vessels, aneurysms (Fig. 38.7).

MICTURATING CYSTOURETHROGRAPHY (MCU)

In this procedure, the dye is injected into the urinary bladder *via* an indwelling catheter and X-rays are taken when patient passes urine.

Indications

1. In children, to demonstrate vesicoureteric reflux
2. Posterior urethral valve
3. Vesical trauma
4. Vesicovaginal or vesicocolic fistula.

Procedure

A catheter is passed into the urinary bladder in a child and the dye is injected. The catheter is removed and the child is screened for vesicoureteric reflux during voiding of urine (Fig. 38.8).

Fig. 38.8: MCU voiding phase: Observe the ureter due to reflux

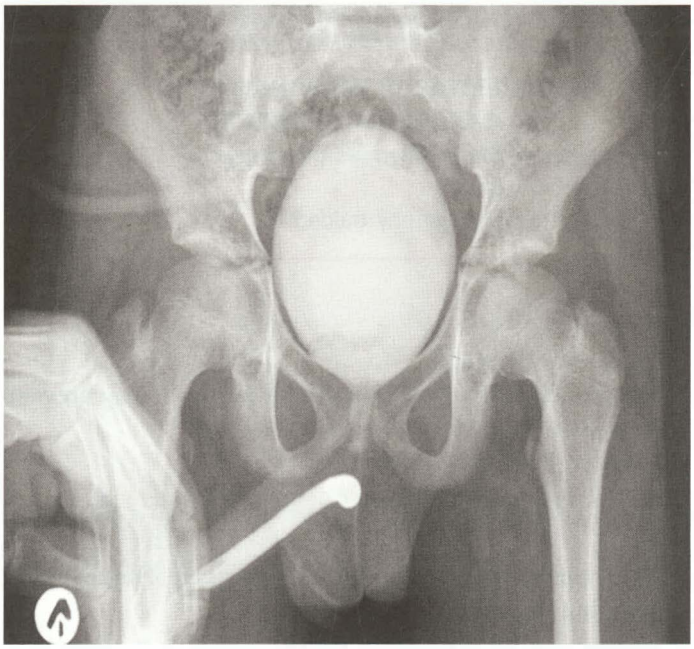


Fig. 38.9: Ascending urethrogram and micturating cysto-urethrogram—stricture urethra

Films

Filling phase, full bladder, voiding and postvoiding phase films are taken.

Newer

Direct or indirect radionuclide cystography wherein isotopes are used. It can pick up minute reflux also.

Complications

Due to the invasive nature of the procedure, urinary tract infection can occur. Hence, prophylactic antibiotics should be used.

ASCENDING URETHROGRAPHY (ASU) OR RETROGRADE URETHROGRAPHY (RUG)

In the diagnosis of urethral stricture, to know the length of stricture, proximal dilatation or diverticulum, urethrography is used (Fig. 38.9).

Indications

- Evaluation of urethral injury
- Investigation of urethral stricture

Contraindication

Urethral haemorrhage

Precaution

Barium and medium containing oil such as lipiodol should not be used because if there is a urethral mucosal tear or breach, it can cause oil embolism. Conray 280 is injected slowly into the urethra.

ULTRASONOGRAPHY (USG) (Fig. 38.10)

- This is a **noninvasive investigation,** using ultrasonic waves (sound waves with frequency > 20,000 Hz). These waves cannot be heard by the human ear but are reflected or absorbed by tissues to various degrees which helps to diagnose different conditions. Ultrasound can be used through different approaches:
- Transabdominal
- Transrectal (Key Box 38.2)
- Transvaginal (used mostly by gynaecologists).

Fig. 38.10: Ultrasound showing hydronephrosis

KEY BOX 38.2

TRANSRECTAL ULTRASONOGRAPHY IN CARCINOMA PROSTATE

- Disruption of the architecture of echoes
- Invasion of capsule
- Biopsy—ultrasonography guided

Limitations of USG

- Operator-dependent
- Air precludes adequate imaging: Bowel gas may prevent satisfactory imaging of pancreas/kidneys.
- Obesity: Poor visualisation.

Uses of USG

A. Fluid can be differentiated from solid tissue. Hence, **cystic swellings** can be made out.

B. **Stones** can be diagnosed. Stones appear as hyperechoic lesions and **postacoustic shadowing.**

C. In an enlarged kidney **thickness of cortex,** disruption of the architecture of echoes can be made out as in hydronephrosis.

D. **Residual urine in the bladder** can be found out, which may be an indication of enlarged prostate. The volume of the prostate can be measured.

Ultrasonography has become the investigation of choice to diagnose foetal hydronephrosis due to various reasons. This has got dual advantages. Firstly, the management of the disease causing hydronephrosis can be planned at the early stage thereby preventing damage to the kidney and secondly intra-uterine interventions are also possible if the need arises.

COMPUTERISED TOMOGRAPHY (CT) SCANNING

- CT can be done with or without contrast (Fig. 38.11).
- CT without contrast (plain CT) is the investigation of choice for evaluation of renal/ureteric colic.
- CT scan can visualise most urinary tract calculi, even radiolucent stones not seen on plain X-ray KUB or IVU.
- Contrast CT gives information about the function also—similar to IVU. However, unlike IVU, which gives information only about the renal parenchyma and collecting system, CT can provide valuable information about perinephric events (e.g. urinoma, abscess, lymph nodes compressing ureters causing hydronephrosis).
- CT angiography is replacing conventional angiography for diagnostic evaluation of renal vascular anatomy.
- It is more useful than **arteriography** to assess and display the images of the body at selected levels.
- It is useful in the diagnosis of kidney tumour and its extent, spread and infiltration.

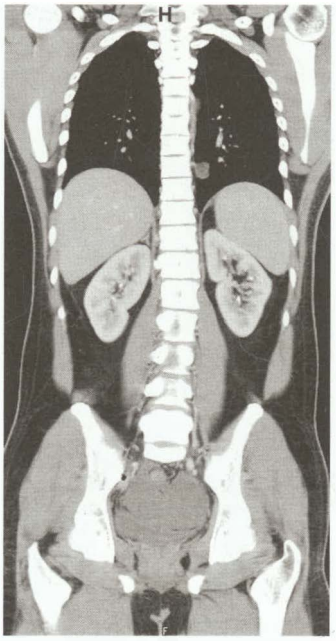


Fig. 38.11: CT scan showing normal kidneys: It gives information about function of kidney. Now more commonly used than IVU

- To stage cancer of prostate, bladder, kidney, testicular tumours and renal trauma (Figs 38.12 and 38.13).

PEARLS OF WISDOM

Presently, CT with contrast is fast replacing IVU for evaluation of urinary tract because of its obvious advantages.

RADIOISOTOPE SCANNING

Gamma camera screening following injection of technetium 99m gives information about proximal tubular function. To assess differential renal functions, diethylene triamine penta-acetic acid (^{99m}Tc DTPA) or dimercapto-succinic acid (^{99m}Tc

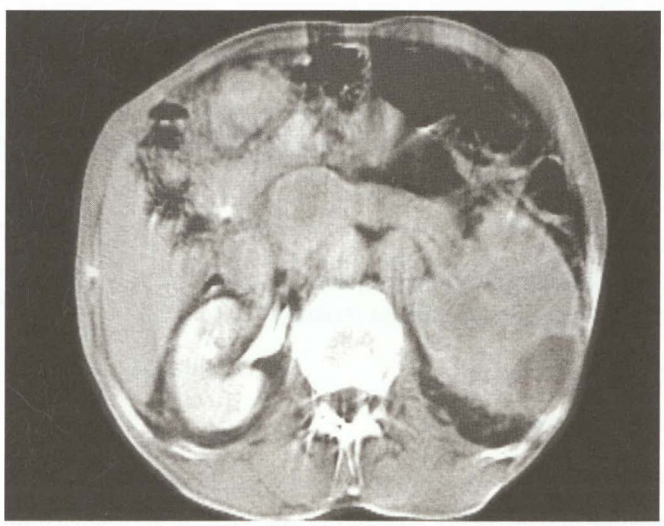


Fig. 38.12: CT scan showing left renal cell carcinoma

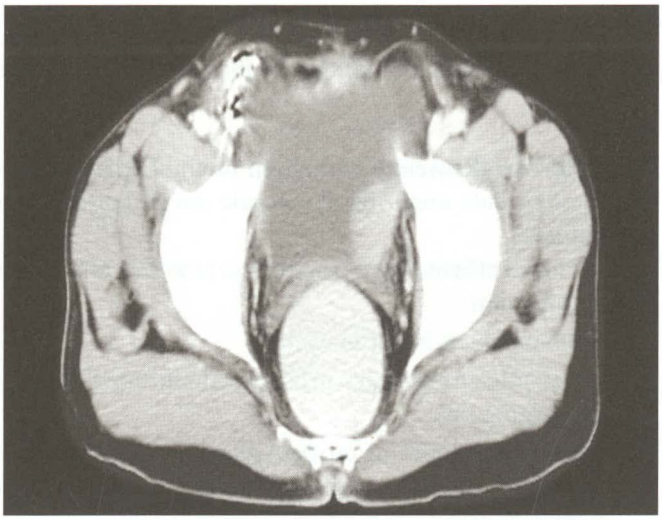


Fig. 38.13: CT scan showing bladder carcinoma

DMSA) is used. It is filtered and secreted into the tubular lumen. Two types are given below.

^{99m}Tc DTPA: Diethylene triamine penta-acetic acid

This scan is done to find out relative functions of both kidneys; it also tells about the total GFR and what percentage of total GFR is contributed by each kidney. Relative function of 45±2% is considered acceptable for each kidney. The main indication for DTPA is long-term hydronephrosis. Examples are newborn with antenatally diagnosed hydronephrosis, children with posterior urethral valves, etc. This scan is also useful in assessing the improvement in relative function of the kidney after surgery for above conditions. The yield of DTPA scan can be improved by injecting IV lasix. This is known as **diuretic renography.** This will unmask marginal pelviureteric junction (PUJ) obstruction (Fig. 38.14).

^{99m}Tc DMSA: Dimercapto-succinic acid

It is used most commonly for cortical imaging. It shows details of renal parenchyma. It is particularly useful when looking for segmental abnormalities of kidney (e.g. renal scarring secondary to conditions such as chronic pyelonephritis and renal tumours—Fig. 38.15 and Key Box 38.3).

ENDOSCOPY

- Cystourethroscopy: Bladder and urethral mucosa can be visualised
- Procedure is done under surface anaesthesia
- Preparation: The external genitalia is cleaned with soap solution or an antiseptic agent and 1% lignocaine jelly is injected into urethra, to provide lubrication and anaesthesia. This should be left in place for 10 minutes for its action.

Uses of cystoscopy

1. Diagnosis of bladder cancer, papilloma, cystitis
2. Position and character of ureteric orifices—in tuberculosis involving the urinary bladder, the ureteric orifices are shifted upwards and gaping—golf hole ureter.

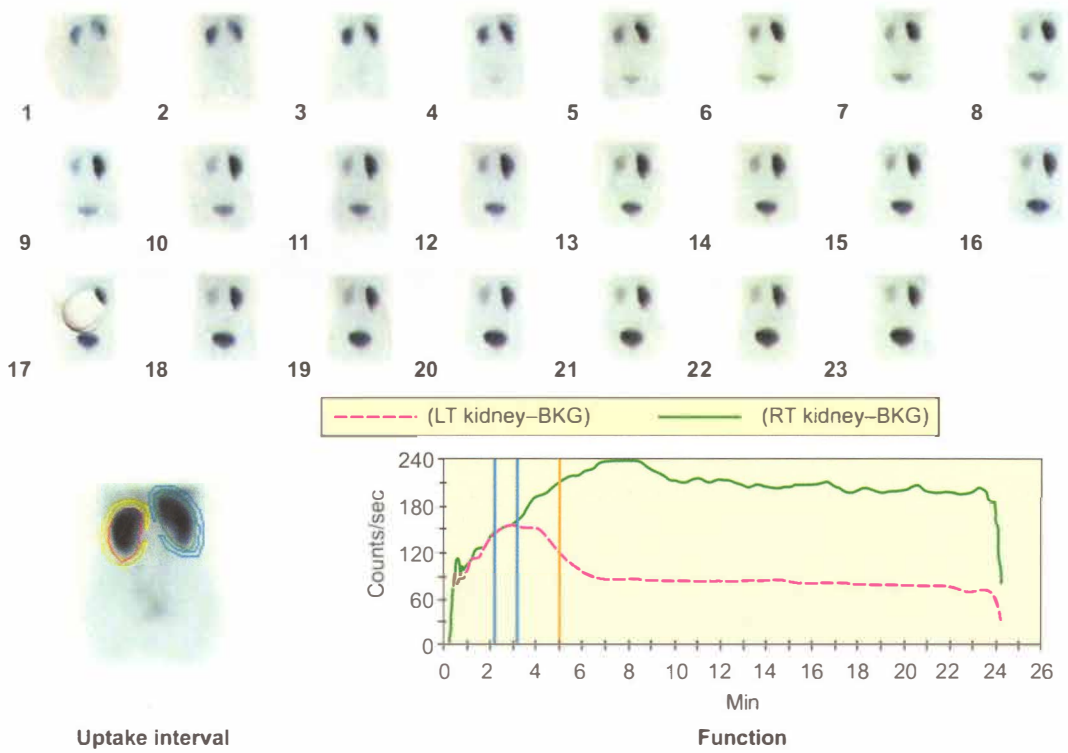


Fig. 38.14: DTPA scan showing sluggish clearance of contrast in the right kidney. Left kidney is normal in clearance and function

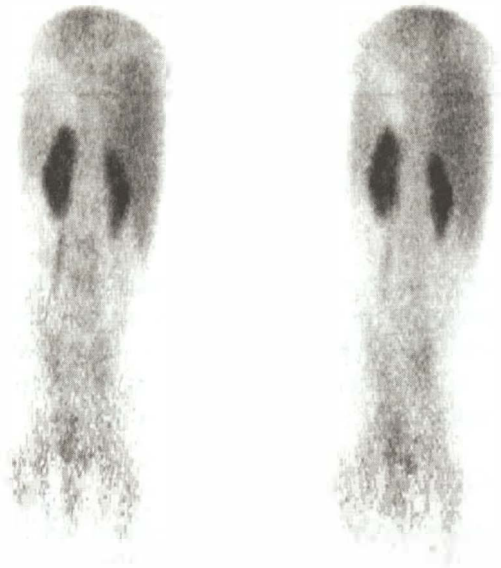


Fig. 38.15: DMSA scan

KEY BOX 38.3

RADIOISOTOPE SCANNING IN UROLOGY

- ^{99m}Tc DTPA – Renal function, drainage
- ^{99m}Tc DMSA – Renal parenchymal images
- ^{99m}Tc Diphosphate – Bone secondaries
- ^{131}I MIBG – Phaeochromocytoma
- ^{99m}Tc Sestamibi – Parathyroid adenoma

3. **Indigocarmine test:** 7 ml of 0.4% dye is injected intravenously. Observe ureteric orifice through cystoscope. Unilateral delay in appearance of dye suggests obstruction. If there is bilateral delay, it indicates impaired renal function.
4. As a preliminary step to do RGP.
5. To rule out involvement of bladder in gynaecological cancer (e.g. cancer cervix).
6. To remove bladder stones
7. For transurethral resection of bladder tumour (TURBT) in early bladder cancers.

URETHROSCOPY

It refers to visualisation of urethra by introducing cystoscope.

Types of urethroscopy

1. **Anterior urethroscopy** is done in urethral stricture or in cases of chronic urethritis. It can rule out strictures due to granuloma.
2. **Posterior urethroscopy:** To visualise prostatic urethra and verumontanum
   - Verumontanum is red in cystitis
   - In chronic prostatitis, prostatic ducts may be seen discharging pus
   - When lateral lobes of prostate gland are enlarged, they project into the internal meatus and produce an inverted 'V' appearance.

MAGNETIC RESONANCE (MR) UROGRAPHY

MRI of the genitourinary system is useful in many situations. It is more expensive compared to other investigations. A few uses have been given below.

Uses of MR urography

- For accurate evaluation of inferior vena cava (IVC) thrombus in renal cell carcinoma (RCC).
- Extrinsic causes of ureteric obstruction causing hydronephrosis (e.g. retroperitoneal fibrosis, pelvic tumours)
- MR urethrography for accurate delineation of urethral injuries.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated.
- Urine examination and CT scan have been discussed in more details.
- MR urography has been added.

MULTIPLE CHOICE QUESTIONS

1. When do you say there is significant haematuria?

A. Presence of more than 2 RBCs/high power field
B. Presence of more than 3 RBCs/high power field
C. Presence of more than 1 RBC/high power field
D. Presence of RBCs in the urine

2. Which one of the following can be detected by urine examination?

A. Transitional cell carcinoma
B. Squamous cell carcinoma
C. Adenocarcinoma
D. Adenosquamous cell carcinoma

3. When do you suspect carcinoma prostate?

A. If patient has urgency of micturition
B. Rectal examination reveal grade 2 enlargement of prostate
C. If prostate specific antigen levels are more than 4 ng/ml
D. If patient has recurrent urinary tract infection

4. Spider leg deformity in intravenous pyelography is a diagnostic sign of which disease?

A. Hydronephrosis B. Polycystic kidney
C. Horseshoe kidney D. Carcinoma kidney

5. Which of the following is a contraindication for intravenous pyelography (IVP)?

A. Staghorn calculi
B. Tuberculosis of kidney
C. Horseshoe kidney
D. Multiple myeloma

6. The following are the advantages of retrograde pyelography over intravenous pyelography (IVP) *except*:

A. Selectively urine sample can be collected from renal pelvis
B. Can be done when the kidney is not visualised by IVP
C. Intravenous contrast need not be given
D. It is an invasive procedure

7. Investigation of choice in foetal hydronephrosis is:

A. CT scan B. MRI scan
C. Ultrasound D. DTPA scan

8. Investigation of choice to know the renal parenchymal/ cortical function or damage is:

A. Intravenous pyelography
B. Contrast enhanced CT scan
C. DTPA scan
D. DMSA scan

9. Investigation of choice to know the renal function/ drainage is:

A. CT scan
B. MRI scan
C. DTPA scan
D. DMSA scan

10. Indigo carmine test is done to study:

A. Ureteric obstruction
B. Prostatic obstruction
C. Urethral obstruction
D. Renal obstruction

ANSWERS

1 B	2 A	3 C	4 B	5 D	6 D	7 C	8 D	9 C	10 A

39

Kidney and Ureter

- Surgical anatomy
- Polycystic kidneys
- Horseshoe kidney
- Renal stones
- Ureteric stone
- Hydronephrosis
- Renal tuberculosis
- Wilms' tumour
- Pyonephrosis
- Perinephric abscess
- Dialysis
- Renal transplantation
- What is new?/Recent advances

SURGICAL ANATOMY OF KIDNEY

- Kidneys are the retroperitoneal organs, two in number on either side of vertebral column. Each kidney is bean or reniform in shape.
  Owing to the presence of liver, right kidney is 1–2 cm lower than the left kidney, extending from L1–L3 and the left kidney extends from T12–L3
- These relationships are very important for surgeons, as the structures mentioned above may get injured during operations on kidney (Table 39.1). Also they may get directly involved by renal malignancies (local spread).

Fascial attachments

Both the kidneys and associated adrenal glands are surrounded by varying degrees of perirenal or perinephric fat and these are enclosed loosely together by the perirenal fascia, commonly called Gerota's fascia.

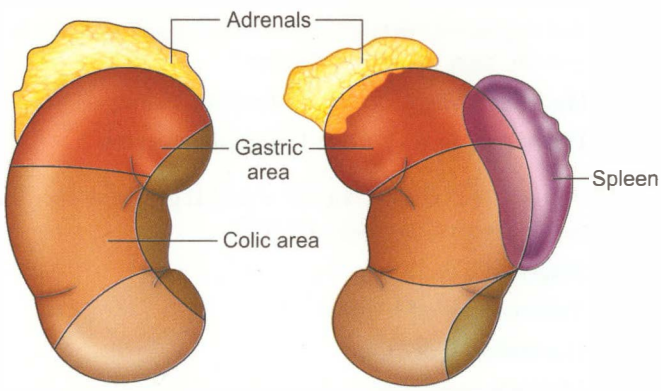


Fig. 39.1: Anterior relationships of the kidney

- Perinephric fat with renal pedicle are responsible for classical ballotability of renal swelling.
- The anterior and posterior sheaths of Gerota's fascia, become fused on three sides around the kidney laterally, medially and superiorly.

Table 39.1 Relations of kidneys

	Right	Left
Anterior (Fig. 39.1)	Below: Hepatic flexure of colon Medial: 2nd part of duodenum	Below: Splenic flexure, pancreas and splenic vessels, below the pancreas is jejunum, above the pancreas is stomach and spleen
Medial	Above: Adrenal, liver, IVC	Above: Adrenal, duodenojejunal flexure, inferior mesenteric vein, ureter
Lateral	Below: Ascending colon	Below: Descending colon
	Above: Liver	Above: Spleen
Posterior	Same in both kidneys. Each kidney rests upon four muscles: Psoas, transversus abdominis, quadratus lumborum, diaphragm	

Superiorly

Gerota's fascia fuses with the diaphragm (as a result ow which kidney moves with respiration). Students should remember here that movement with respiration is a characteristic feature of intraperitoneal organs and masses, and many times this feature is used to support the diagnosis of intraperitoneal organ. However, even though the kidney is a retroperitoneal organ, it also moves with respiration.

Medially

Gerota's fascia of one side crosses the midline and fuses with Gerota's fascia of the opposite side.

Inferiorly

Gerota's fascia remains an open potential space containing ureter and gonadal vessels.

- Gerota's fascia forms an important anatomical barrier and tends to confine pathological processes originating from the kidney.
- However, because of its deficiency inferiorly, a collection within Gerota's fascia may track down and extend into pelvis.

Some points to remember

- Renal artery is an **end artery** and the entire renal arterial system is composed of end-arteries (without anastomosis and collateral circulation). Occlusion of any branches of renal artery (within kidney known as **segmental arteries**) results in **infarction** of area supplied by it.
- In contrast, the renal parenchymal **veins anastomose freely** with each other and even with perinephric veins.

POLYCYSTIC KIDNEYS (CONGENITAL CYSTIC KIDNEYS)

- This is an autosomal dominant disease transmitted through chromosomes from anyone of the parents. Early onset congenital cystic kidneys are found to be autosomal recessive.
- More common in women

Types

- **ADPKD: A**utosomal **D**ominant **P**olycystic **K**idney **D**isease (Figs 39.2 and 39.3).
- **ARPKD: A**utosomal **R**ecessive **P**olycystic **K**idney **D**isease.

Associated lesions

- Congenital polycystic disease of liver (18%).
- Congenital polycystic disease of pancreas.
- Congenital polycystic disease of ovary or testis.
- Berry aneurysms

Fig. 39.2: Autosomal dominant polycystic kidney disease

Pathology

- During development, some of the uriniferous tubules fail to join with the collecting ducts. Such uriniferous tubules develop into cysts. The important pathological features are as follows:
- Both kidneys are affected (Fig. 39.2)
- They enlarge often to 3–4 times the normal size.
- Cysts are distributed evenly throughout cortex and medulla.
  - Content of cyst varies but it is not urine.
- The kidneys are studded with **multiple large cysts**
- When the cyst ruptures into the pelvis of the kidney, it results in **haematuria**.
- As the disease progresses, cysts progress in size which lead to **pressure atrophy** of the functional renal parenchyma leading to **renal failure**.

Types (for recessive)

1. **Foetal type:** Autosomal recessive disease may cause intrauterine death due to obstructed labour.
2. **Infantile type:** Autosomal recessive, early renal failure with death within 3–5 years of life.
3. **Adult type:** Autosomal dominant type. Presents in the 3rd or 4th decade with symptoms (Fig. 39.3).

Clinical features

- Even though congenital, it manifests at middle age around 40 years.

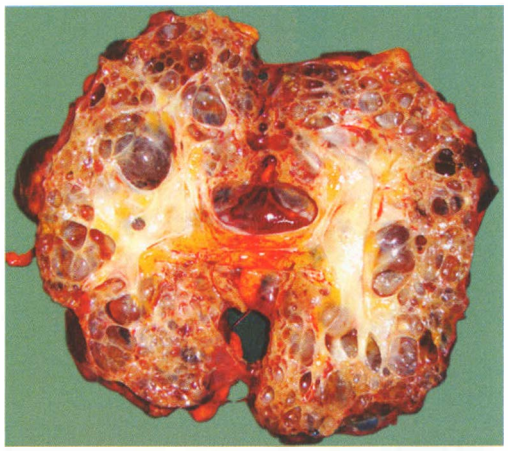


Fig. 39.3: Autosomal dominant polycystic kidney disease: Cut section

- Dull aching pain in both loins is due to stretching of the renal capsule (dragging pain).
- Microscopic or macroscopic **haematuria** in 70–80% cases.
- **Hypertension** (secondary) (75%) is due to renal ischaemia which stimulates juxtaglomerular apparatus to secrete renin. It may also be related to a separate genetic factor.
- **Bilateral renal mass:** Both kidneys are enlarged, surface is nodular or bosselated, firm to hard and some times cystic.
- Features of **renal failure:** Thirst, vomiting, abdominal distension due to paralytic ileus, anuria, uraemic smell, coated tongue, anaemia.
- Infection, pyelonephritis
- Acute pain: If there is haemorrhage into or infection of a cyst. Colicky pain is due to blood clot in the ureter.

Diagnosis

1. Serum urea and creatinine to rule out renal failure. Normal creatinine levels: 0.8–1.6 mg%. Normal urea: 20 to 40 mg%.

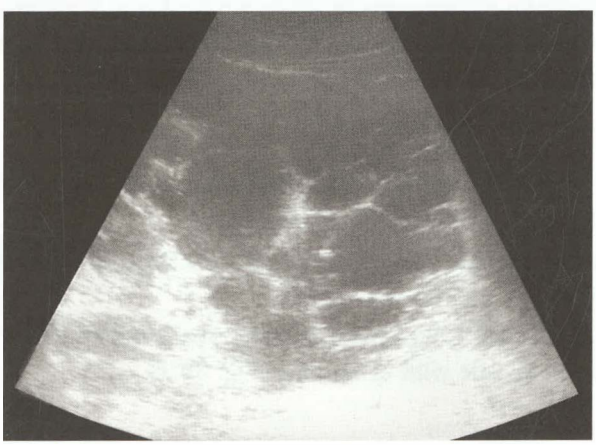


Fig.39.4: USG of autosomal dominant polycystic kidney disease

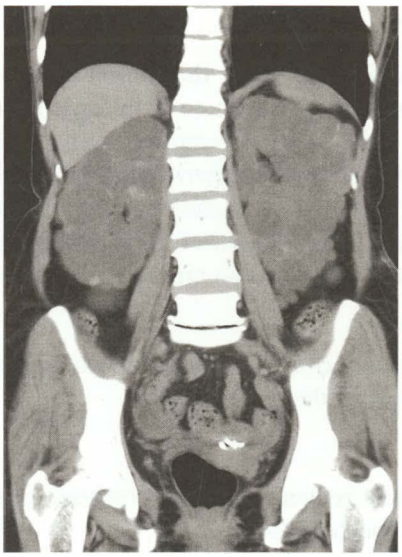


Fig. 39.5: CT of autosomal dominant polycystic kidney disease

2. Plain X-ray KUB: Enlarged kidney can be seen because of changes in the density between the perirenal pad of fat and the kidney.
3. Abdominal USG/CT scan to confirm the diagnosis (Figs 39.4 and 39.5).
4. IVU: The **spider leg deformity** of the calyces.

Treatment

- **Asymptomatic** polycystic kidney does not require any treatment other than **regular follow-up**.
- **Polycystic kidney with hypertension:** Control of hypertension with drugs. When hypertension becomes uncontrollable, bilateral nephrectomy followed by renal transplantation should be done.

CLINICAL NOTES

My MS Exam Case, June 1986, Wenlock Hospital, KMC, Mangalore.

A 45-year-old lady with mass abdomen was allotted as my long case. She had undergone open cholecystectomy 3 months back (ultrasound was not available at that time). She had a vague renal mass in the right loin. I was not sure. When I recorded BP it was 200/110 mm of Hg. I carefully palpated the opposite side where I was able to palpate a renal mass. The diagnosis became evident. It was polycystic kidney. In fact one of the examiners was the surgeon who had operated on this patient for open cholecystectomy. During that surgery, he discovered enlarged kidneys. The patient was asymptomatic.

- **Infected cyst or if pyelonephritis** develops: Appropriate antibiotics are given and if necessary, the cyst should be aspirated using ultrasound guidance.
- **Polycystic disease with renal failure**, emergency dialysis followed by renal transplantation is the treatment of choice. The related donor should be screened for polycystic trait.

HORSESHOE KIDNEY

- During development, 2 mesonephric buds appear on the side of the future vertebral column and grows into metanephros. Mesonephric buds form ureter and metanephros kidneys.
- **If fusion occurs at lower pole**, it results in a classical **horseshoe kidney** (Fig. 39.6).
- Rarely, upper polar fusion can occur giving rise to reverse horseshoe kidney.
- Inferior mesenteric artery crosses the isthmus at the level of L3–L4. Hence, horseshoe kidney cannot ascend. It is felt lower down in the abdomen.

Clinical features

- It can be asymptomatic for many years.
- A palpable mass below and to the right and to the left of umbilicus or umbilical region can be a horseshoe kidney.
- **Recurrent urinary tract infection (UTI)** is common because the ureters are angulated over the kidney isthmus.
- They are more **prone for hydronephrosis** due to the angulation of ureters.

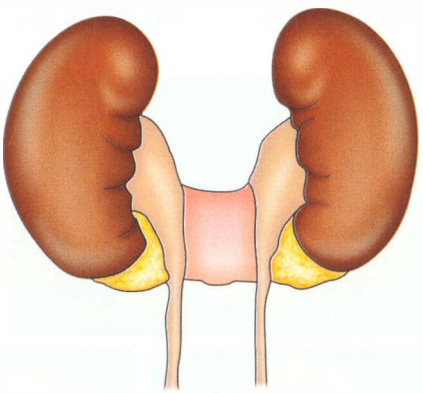


Fig. 39.6: Horseshoe kidney

KEY BOX 39.1

ASSOCIATED ANOMALIES

- Spina bifida
- Congenital hemivertebra
- Turner's syndrome
- Cleft lip and cleft palate

- **Rovsing's sign:** Hyperextension of the spine results in abdominal pain, nausea or vomiting due to stretching of the capsule.

Diagnosis

1. Ultrasonography (USG), to locate the kidney.
2. IVU: Upper and middle calyx are directed laterally but **lower calyx is directed medially** where there is fusion, which is characteristic of horseshoe kidney.
3. CT scan or isotope renogram are confirmatory.

Treatment

- Indicated only when there are complications.
- Removal of the stone, or repair and reconstruction of the hydronephrosis are done in the usual manner.

RENAL STONES

Aetiopathogenesis

1. **Infection:** Organisms such as Proteus, Pseudomonas, Klebsiella produce recurrent UTI. These organisms produce urea, cause stasis of urine and precipitate stone formation. Nucleus of the stone may harbour these bacteriae (Fig. 39.7).
2. **Hot climates** cause increase in concentration of solutes, resulting in precipitation of calcium and formation of calcium oxalate stones.
3. **Dietary factors**
   - Diet rich in red meat, fish, eggs can give rise to aciduria.
   - Diet rich in calcium—tomatoes, milk, spinach, rhubarb produce calcium oxalate stones.
   - Diet lacking in vitamin A causes desquamation of renal epithelium which precipitates calcium, alters it and forms stones.

   Epitaxy: Growth of one type of stone over another type.

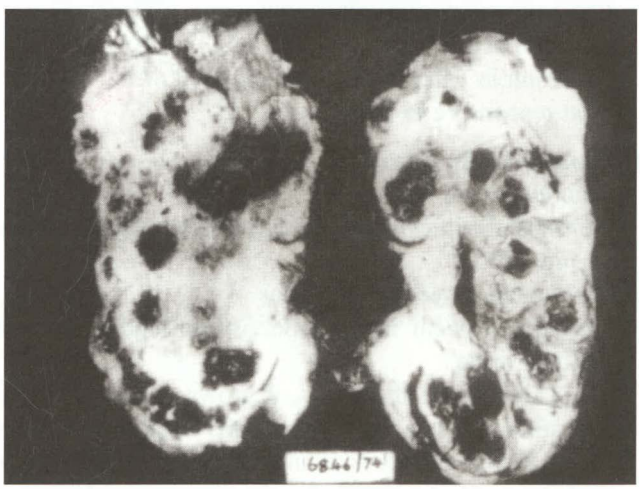


Fig. 39.7: Chronic pyelonephritis with calculi (*Courtesy:* Prof Sasidharan, Head, Dept of Urology (2002–2008), KMC, Manipal)

4. Metabolic causes
- Hyperparathyroidism increases serum calcium levels resulting in hypercalcinosis and pelvic stones.
- Gout increases uric acid levels and causes multiple uric acid stones.

5. Immobilisation: Paraplegic patients secrete large amounts of calcium in the urine resulting in calcium oxalate stones (they pass skeletons in urine).

6. Decreased urinary citrate: Citric acid (300–900 mg/24 hours) keeps the urinary pH low. When citric acid levels decrease, it promotes precipitation of urinary calcium. Citrate excretion is under hormonal control.

7. Inadequate urinary drainage as in cases of horseshoe kidney, unascended kidneys are more vulnerable for development of stones due to stasis.

8. Randall's plaques: Randall has suggested that initially a small erosion or an ulcer develops at the tip of renal papilla on which minute concretions or minor calcium particles get deposited and give rise to stone formation.

Types of renal stones

1. Calcium oxalate stone
- Called as mulberry calculi
- Common type of stone
- It is irregular having sharp projections
- Oxalate stone is hard and single
- Produces haematuria very early, resulting in deposition of blood over the stone giving a dark colour to the stone.
- It occurs in infected urine (Fig. 39.8)
- Contains alternate layer of calcium and bacterial vegetation. It is visualised in plain X-ray KUB.

2. Uric acid stone (Fig. 39.10)
- Multiple, small, hexagonal, faceted, yellow coloured.
- Contain calcium oxalate which makes them opaque. Pure uric acid stones are radiolucent.
- Occur in acidic urine
- Common in patients who consume red meat
- Best responsive to lithotripsy

3. Phosphate stone (Figs 39.11 to 39.13)
- Smooth, round
- Consists of triple phosphate of calcium, magnesium and ammonium.
- Dirty white to yellow in colour
- Commonly occur in renal pelvis and tend to grow in alkaline urine.
- As it enlarges in the pelvis, it grows within the major and minor calyces and slowly forms **staghorn calculus**. This calculus produces recurrent urinary tract infection and haematuria and slowly damages the renal parenchyma (Fig. 39.9).

4. Cystine calculus
- Cystinuria is an inborn error of metabolism which occurs due to decreased resorption of cystine from the renal tubules.
- Occurs in young girls at puberty
- Increased excretion of cystine in urine results in cystine calculus.
- Stones are hard and radio-opaque due to sulphur.

Clinical features

- **Renal pain:** Dull aching to pricking type of pain posteriorly in the renal angle formed by the sacrospinalis and 12th rib. Murphy's kidney punch test demonstrates tenderness at renal angle. The same pain may some times be felt anteriorly

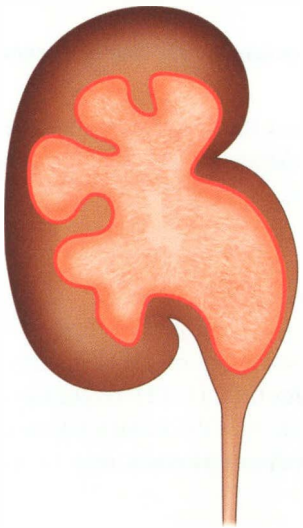


Fig. 39.9: Staghorn calculi

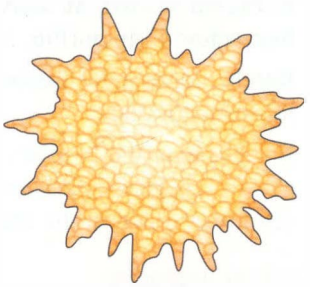


Fig. 39.8: Oxalate stone

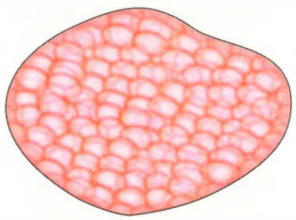


Fig. 39.10: Uric acid stone

Figs 39.11 to 39.13: Staghorn calculi and phosphate calculi

in the costal margin. Hence, it is described as costovertebral pain. Nausea and vomiting is due to intense sympathetic stimulation caused by stretching of renal capsule mediated by coeliac plexus.

- **Ureteric colic:** When the stone is impacted in the pelviureteric junction or anywhere in the ureter, it results in severe colicky pain originating at the loin and radiating to the groin, testicles, vulva and medial side of the thigh. This may be associated with strangury. The referred pain is due to irritation of the genitofemoral nerve.
- **Haematuria** is common with renal stone because the majority of stones are oxalate stones. The quantity of blood lost is small but it is fresh blood (Key Box 39.2).
- **Recurrent UTI:** Fever with chills and rigors, burning micturition, pyuria may occur, along with increased frequency of micturition.
- **Guarding and rigidity** of the back and abdominal muscles during severe attack of pain.

Complications

1. **Calculous hydronephrosis** occurs due to back pressure producing renal enlargement. Stretching of the renal capsule results in pain. In such cases, an associated palpable kidney mass suggests hydronephrosis.
2. **Calculous pyonephrosis:** Infected hydronephrosis where in the kidney is converted into a bag of pus.
3. **Renal failure:** Bilateral staghorn stones may not be symptomatic until they present with uraemia and renal failure.
4. **Squamous cell carcinoma:** Long-standing stones increase the risk of carcinoma.

Investigations

1. **Blood urea and creatinine to rule out renal failure.**
2. **Plain X-ray KUB** (Figs 39.14 and 39.15)
   - To diagnose stones 90% of the renal stones are radio-opaque.
   - Enlarged renal shadow can be seen.
3. **USG**
   - Presence of the stone can be diagnosis
   - Exact size and location of the stone can be evaluated.

KEY BOX 39.2

CAUSES OF HAEMATURIA: RENAL CONDITIONS

1. Polycystic disease of kidney
2. Renal stone, ureteric stone
3. Renal tuberculosis
4. Carcinoma kidney
5. Papilloma of kidney
6. Renal infarction

Fig. 39.14: Plain X-ray KUB showing large stone in the pelvis

Fig. 39.15: Plain X-ray KUB showing bilateral staghorn calculus

4. **IVP**
   - To locate the stone exactly in relation to kidney and ureter and to assess renal function. A nonradiopaque stone can be seen as a filling defect. Hydronephrosis and hydronephroureterosis can also be seen.
   - Now noncontrast CT scan and contrast CT scan are used for more accurate detection of causes of abdominal colic.
5. **Urine for culture and sensitivity.**

Treatment

The treatment of renal stones can be divided into nonoperative treatment and operative treatment.

I. Nonoperative treatment

1. **Conservative:** Small stones less than 5 mm in size pass off with intake of copious amount of fluids and at times forced diuresis. Intravenous hydration followed by intravenous frusemide may help pass the stones spontaneously.
2. **Extracorporeal shock wave lithotripsy (ESWL):** After cystoscopy, a ureteric stent (Double J stent) is placed into the ureter on the side of a large renal stone (Fig. 39.16). Shock waves are generated (around 500–1500 shock waves) which blast the stone. The stones get crushed and most of the stones will come out by the side of the stent (Key Box 39.3). Small stones can be removed without prior crushing.

Steinstrasse

- It means "stone street"
- It is a condition which follows the use of ESWL.
- Small pieces of fragmented calculi collect and obstruct in the distal ureter.

Laser lithotripsy

- Holmium-YAG laser (Ho-YAG) is the most effective with good margin of safety.
- Wavelength 2100 nm
- Ho-YAG laser lithotripsy occurs primarily through a photothermal mechanism that causes stone vaporisation.
- They have the ability to fragment all types of stones regardless of the composition.

ESWL is not effective in fragmenting **cysteine stones** because of its crystal lattice.

II. Operative treatment

1. Endoscopic procedures
2. Open procedures
3. Special situations

1. *Endoscopic procedures*

Percutaneous nephrolithotomy (PCNL): Retrograde pyelography (RGP) is done when the stone is located in the pelvis of the kidney. With a small 1 cm incision in the loin, the PCN needle is passed into the pelvis of the kidney and is confirmed by fluoroscopy. A guide wire is passed through the needle into the pelvis of the kidney. The needle is withdrawn, with the guide wire left within the pelvis. Dilators are passed over the guide wire and a working sheath is introduced into the pelvis. A nephroscope is passed into the pelvis and if the stone is small, it can be taken out. If it is big, it may have to be crushed using ultrasound probes and the fragments are removed. Ultrasound or pneumatic energy is used for fragmenting.

Complications of PCNL

- Injury to the colon, sepsis
- Injury to the blood vessels
- Urinary leak may persist for a few days.

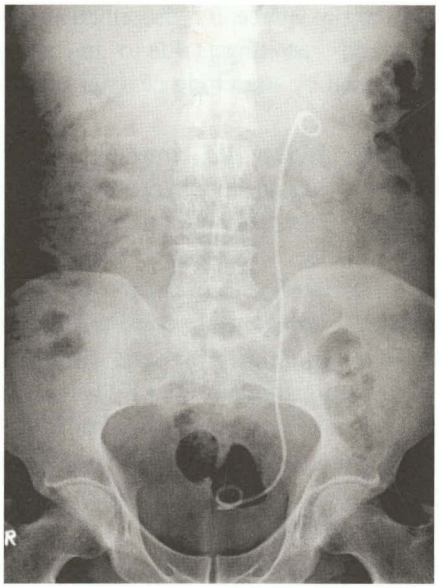


Fig. 39.16: After clearance, DJ stent in place

2. *Open surgical procedures*

Depending upon the location of the stone, various types of procedures are done. They are as follows:

A. Pyelolithotomy: When there is extrarenal pelvis.

B. Nephrolithotomy: When there is intrarenal pelvis, the stone has to be taken out through the kidney parenchyma.

C. Extended pyelolithotomy: By retracting the kidney parenchyma laterally, the incision over the pelvis can be extended over to the calyx and the stone can be extracted from the calyx. Even a large staghorn calculus can thus be removed.

D. Pyelonephrolithotomy: Stone is extracted through an incision in the pelvis as well as the renal parenchyma.

E. Partial nephrectomy: When the stone from a lowermost calyx is impacted, a lower pole nephrectomy can be done.

F. Nephrectomy: When the kidney is destroyed by pyonephrosis, following obstruction by stone.

3. *Special situations*

A. **Bilateral renal stones:** Kidney with better function has to be operated first. 1–2 weeks later, the opposite side can be operated.

B. If there is pyonephrosis with a severe degree of fever, pain and tenderness, nephrostomy is done and a tube drain is

KEY BOX 39.3

ESWL

Advantages	Disadvantages
• No incision	• Cost factor
• No pain	• Availability

placed in the pelvis of the kidney for drainage of pus and urine. Once the pus is cleared, fresh assessment of renal function is done. If the kidney is nonfunctioning, nephrectomy is done. If the kidney is functioning ESWL/PCNL/open procedure is done. This is known as percutaneous nephrostomy (PCN).

PEARLS OF WISDOM

Open procedures for the management of stone disease have become obsolete and are found in old surgery textbooks.

URETERIC STONE

Stones come down from pelvis of the kidney and may get impacted at any site of anatomical narrowing of ureter, namely:

1. Pelviureteric junction
2. Crossing of the iliac artery
3. Crossing of the vas deferens or broad ligament.
4. Site of entry into the bladder wall
5. Ureteric orifice

This may lead to hydroureteronephrosis, renal parenchymal atrophy, infection and pyonephrosis.

Clinical features

- Pain in the loin radiating to groin: Pain is severe, colicky, intolerable and lasts for a few hours. When stone descends into lower ureter, pain radiates to the testicles, labia majora and to the upper portion of thigh due to irritation of genitofemoral nerve. Colic lasts for about 4–6 hours and is relieved by antispasmodics, narcotics and NSAID.
- An attack of haematuria or pyuria
- Guarding and rigidity of the abdominal wall if present on the right side, is confused with acute appendicitis.

Investigations

Same as renal stone

Treatment

1. Most of the ureteric stones pass *via naturalis* (urine). The patient is asked to consume a lot of water and antispasmodics.
2. **Flushing therapy:** About 2 L of IV fluid, with 20–40 mg inj frusemide (Lasix). It can be repeated for a few days.
3. **Stone in the upper ureter:** ESWL is the ideal treatment.

   Retrograde intrarenal surgery (RIRS) for ureteric, renal and calyceal stones. Flexible ureteroscopy with laser fulguration can also be done.
4. **Middle ureteric stone:** ESWL, ureteroscopy basketing or open surgery (ureterolithotomy).
5. **Lower ureteric stone:** Ureteroscopic removal. With the usage of ureteroscope passed through the urethra, direct visualisation and manipulation of the stone even if it is impacted can be done. A laser lithotripter or ultrasonic lithotripter can be used to disintegrate the stone.
6. **Vesicoureteric junction:** Ureteroscopic removal or endoscopic meatotomy of vesicoureteric junction. For a stone impacted at ureterovesical junction, cystoscopy is performed. Ureteric orifice is identified and a cut is given at its mouth. Under fluoroscopic monitoring, the stone can be manipulated and basketed out using a dormia basket or other types of baskets available.
7. **An impacted stone** which is not amenable to ESWL, fluoroscopic or ureteroscopic manipulation have to be extracted by ureterolithotomy (open surgical method).

Prevention of stone disease

1. Metabolic work up of urine and blood for identifying metabolic causes. Example: Hyperparathyroidism to be ruled out by 24 hours urine analysis for calcium, phosphate and uric acid levels.
2. Fluid management: 1.5 L/day
3. Dietary adjustments: Red meat to be avoided (rich in uric acid).
4. Drug treatment
   - Xyloric, sodium bicarbonate: Uric acid stones
   - Potassium citrate: Calcium stones
   - Thiazides small dose: Calcium stones
   - D-penicillamine: Cystine stones
   - Protease inhibitors: Infection stones against *E.coli*
5. Ultrasound to be done once in 6 months.

HYDRONEPHROSIS

Definition

Aseptic dilatation of the whole or part of the pelvicalyceal system of the kidney due to partial or intermittent interruption to the outflow of urine.

Causes of unilateral hydronephrosis

I. Intraluminal (within the lumen)

- Stones and papillary necrosis in diabetes mellitus, analgesic nephropathy.

II. Intramural (in the wall)

1. Congenital

A. Pelviureteric junction (PUJ) abnormality (PUJ dyskinesia or achalasia of PUJ) is a congenital lesion where hydronephrosis occurs due to failure of transmission of neuromuscular impulses through the narrow PUJ. It can

also be bilateral. Male : Female ratio is 2 : 1. More common in males on the left side.

B. Ureterocoele, congenital narrow ureteric orifice.

2. Acquired

A. Carcinoma of the ureter or carcinoma of the bladder involving the ureteric orifice.

B. Stricture of the ureter secondary to stone: After dislodgement of the stone there can be inflammatory stricture of the ureter.

C. Tuberculosis of the ureter and bladder.

III. Extramural

1. Involvement of ureter by carcinoma cervix, rectum, bladder, the retroperitoneal tumours, primary or secondary deposits in lymph nodes.

2. **Obstruction by aberrant vessels** (Fig. 39.18)

- Aberrant renal artery going to the lower pole of kidney can cause obstruction to the ureter causing hydronephrosis.

3. Retrocaval ureter

4. Horseshoe kidney

Causes of bilateral hydronephrosis

Lower urinary tract obstruction below the level of bladder neck will give rise to bilateral hydronephrosis (Ormond's disease). Any of the causes for unilateral hydronephrosis when present bilaterally cause bilateral hydronephrosis (Fig. 39.17).

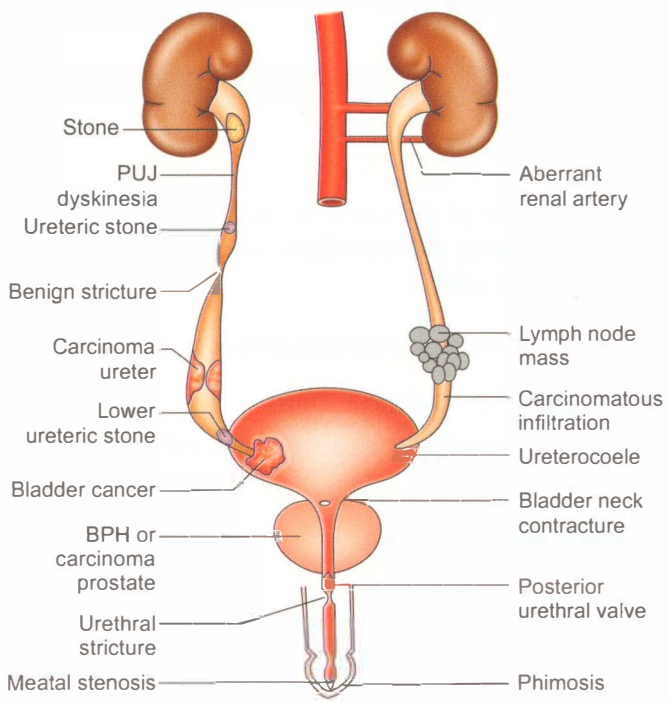


Fig. 39.17: Causes of hydronephrosis

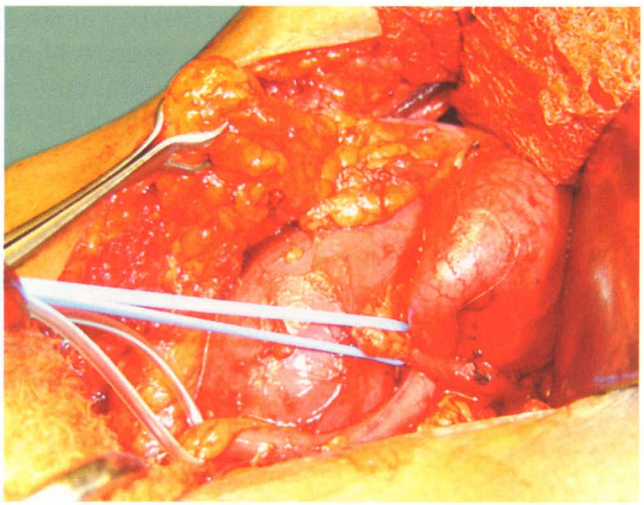


Fig. 39.18: PUJ obstruction with aberrant crossing vessel

I. Causes in children

1. Phimosis
2. Meatal stenosis
3. Posterior urethral valve
4. Bilateral vesicoureteric reflux

II. In young adults

1. **Stricture urethra** is commonly due to gonococcal urethritis. Iatrogenic strictures following instrumentation of urethra or following a rupture urethra later is becoming more common.
2. **Bilateral aberrant vessels:** Often a time, these may be the branches of renal artery and vein which cross the ureters.

III. Causes in the middle age and above

1. Benign prostatic hypertrophy (BPH)
   - Common cause in middle age
2. Contracture of bladder neck
3. Idiopathic retroperitoneal fibrosis (Ormond's disease).

IV. Physiological: Pregnancy

Due to growing foetus and partly due to the hormone progesterone.

Pathogenesis

The back pressure effect depends upon the type of the pelvis (Figs 39.19 and 39.20).

1. In patients with intrarenal pelvis the kidney gets damaged very early. As the time goes on, the urine gets diluted. All the salts are absorbed and is replaced by a watery type of fluid having a specific gravity of 1010.
2. Patients with extrarenal pelvis have minimal damage to the renal parenchyma for a long time.
3. Even if there is complete obstruction to the urinary flow in a hydronephrotic kidney, some amount of urine is secreted, some of it is absorbed by the renal pelvis and the collecting

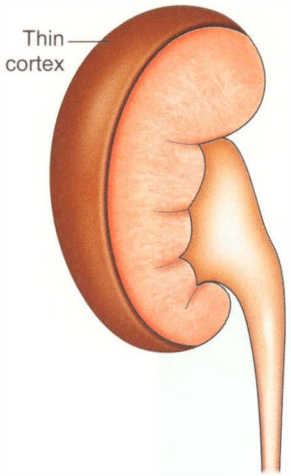


Fig. 39.19: Kidney with intrarenal pelvis

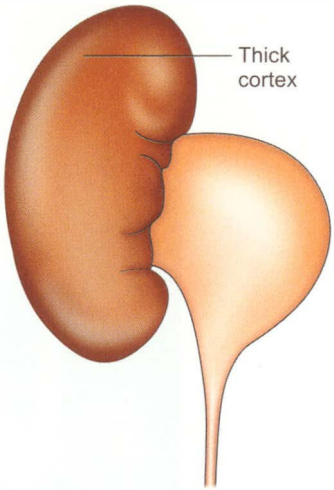


Fig. 39.20: Kidney with extrarenal pelvis

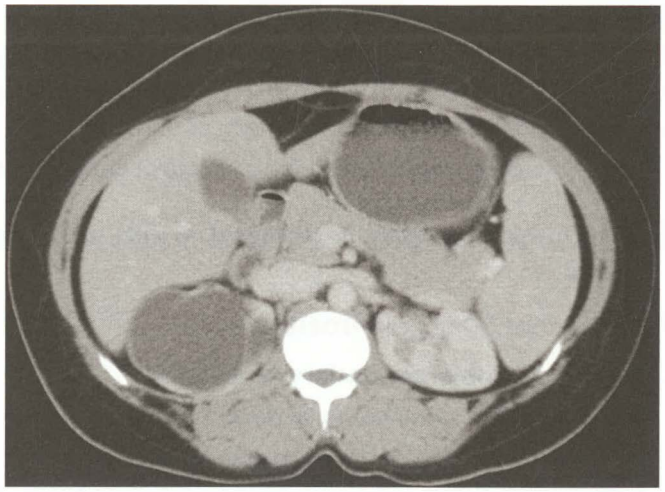


Fig. 39.21: Right hydronephrosis—CT picture. Left kidney normal

tubule and some by the lymphatics of the interstitial tissue of the kidney. The urine is thought to enter the interstitial space of the kidney from the pelvis through microscopic discontinuity **(pyelo-sinus backflow-break)** in the covering epithelium.

- If the disease progresses further, it leads to a nonfunctional kidney.
- If the disease is bilateral, it may give rise to uraemia.

Clinical features

1. Painless enlargement of the kidney. A renal mass is felt in the loin with a smooth surface and firm in consistency (tensely cystic).
2. A dull-aching pain in the loin
3. Previous history of calculus disease
4. Hypertension and haematuria are rarely seen in hydronephrosis.
5. **Dietl's crisis:**
   - It is intermittent hydronephrosis
   - This is common in calculous hydronephrosis.
   - Following an attack of renal colic, ureteric obstruction occurs due to stone which results in enlargement of the pelvis of the kidney resulting in a palpable mass in the loin. After a few hours, the mass disappears due to the passage of a large quantity of urine due to reflux polyuria or due to slipping of the stone.
6. The symptoms of primary cause may be evident in the history, e.g. colicky radiating abdominal pain due to stones and haematuria.

Investigations

1. Plain X-ray KUB

- Enlarged renal outline can be made out
- Demonstration of stone

2. **USG** can detect enlarged kidneys and can detect the cause for hydronephrosis in majority of cases.
3. **CT scan** is the investigation of choice. It can assess anatomy and function more accurately than IVU (Fig. 39.21).

4. **Intravenous pyelography (IVP)** (Fig. 39.22)
   - Normally calyces are concave.
   - They become flat and later convex/club shaped followed by dilatation of pelvis and ureter depending upon the level of obstruction.
   - In hydronephrosis with gross impairment of renal function, the dye may not be visible for a few hours in the X-ray. In such cases, large quantity of dye (100–200 ml) may have to be used and the pictures may have to be taken even after 24 hours. Such a situation is seen in PUJ dysfunction.
5. **Isotope renography:** Technetium 99m-labelled DTPA (diethylene triamine penta-acetic acid) scan using a Gamma camera. The above Gamma radiation emitter is injected intravenously and it can be detected to have been trapped in the urinary tract above the level of obstruction. It does not get washed off even after giving frusemide injection.

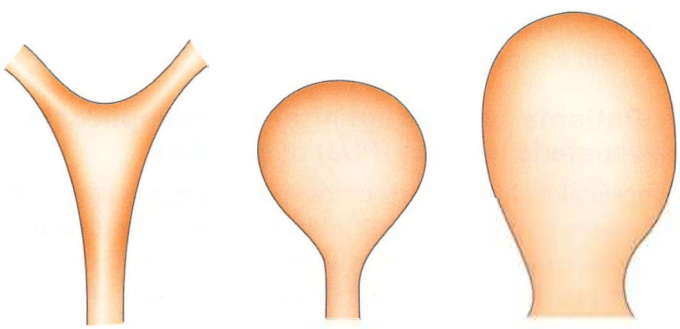


Fig. 39.22: Normal, flattened and club-shaped calyx

6. Retrograde pyelography (RGP)

- When the IVP fails to demonstrate the kidneys, RGP is a useful investigation. It can be done just prior to definitive surgery for confirmation of the site of obstruction.
- Less quantity of dye is required and better configuration of calyces can be made out.

7. Blood urea, creatinine is estimated to rule out renal failure

Treatment of hydronephrosis

I. Hydronephrosis secondary to a cause

Treatment of the cause has to be done. Examples:

a. **Stones:** Pyelolithotomy, ureterolithotomy

b. **Stricture:** Strictureplasty or excision and end-to-end anastomosis.

c. **Aberrant vessel:** Transection of the ureter and anastomosis in front of the vessel.

d. **Phimosis:** Circumcision

e. **Meatal stenosis:** Meatoplasty

f. **Posterior urethral valve (PUV):** Transurethral fulguration of the valve.

g. **Benign prostatic hypertrophy (BPH):** Transurethral resection of the prostate (TURP).

h. **Carcinoma of prostate:** TURP + bilateral orchidectomy + hormonal therapy.

i. **Stricture urethra:** Visual internal urethrotomy or urethroplasty.

Principles of surgery

1. **Nonfunctioning kidney** with thinned out cortex and hydronephrosis/pyonephrosis—nephrectomy.
2. If the cortical thickness is adequate (0.5 cm) by ultrasonography, even though it is a nonfunctioning kidney, a **preliminary nephrostomy** to decompress the system has to be done. Reassessment of the renal function is done after a few days. If the renal function improves, definitive surgery for hydronephrosis can be done. If it remains a nonfunctioning kidney and the opposite kidney is normal, nephrectomy is done.
3. In B/L hydronephrosis, kidney which is functioning better should be operated first.

II. Patients with congenital hydronephrosis—pelviureteric junction (PUJ) dysfunction

Congenital hydronephrosis needs special mention here. With increasing use of obstetric ultrasound, the incidence of antenatally detected foetal hydronephrosis is on the rise. In the present scenario, antenatal detection of foetal hydronephrosis has become the most common mode of presentation of congenital hydronephrosis.

Fig. 39.23: Gross hydronephrosis due to PUJ obstruction

Congenital hydronephrosis is defined as the antero-posterior diameter of renal pelvis > 10 mm at > 20 weeks of gestation. PUJ obstruction is the main cause (Fig. 39.23). These foetuses undergo serial ultrasound monitoring during the rest of pregnancy and based on the increase or decrease in the pelvic diameter during this period, postnatal management can be planned even before the child is born. Recently one study has graded antenatal hydronephrosis due to PUJ on the basis of pelvic diameter and proposed the management guidelines (Dr Vikas Jain, Professor Sasidharan et al—KMC, Manipal).

INDICATIONS FOR SURGERY (Key Box 39.4)

KEY BOX 39.4

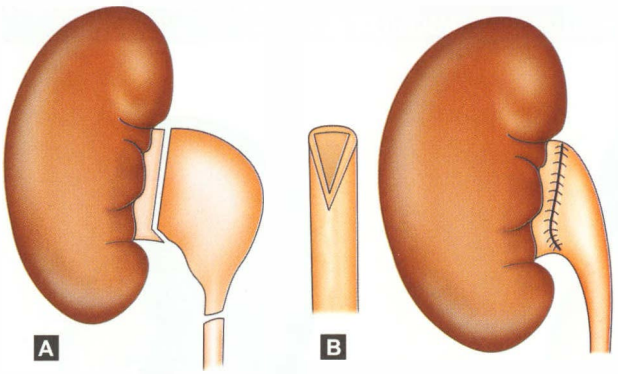
INDICATIONS FOR SURGERY

Pain
Atrophy of kidney (damaged)
Infection
Nephrosis—hydronephrosis
Remembered as **PAIN**

Grades of renal pelvic diameter and management

I	Mild	11–20 mm
II	Moderate	21–35 mm
III	Severe	>35 mm

- **Grade I hydronephrosis** can be managed conservatively by serial monitoring of pelvic diameter by ultrasound and renal functions by DTPA scan. This is known as conservative or nonoperative management. These kidneys improve over a period of time.
- **Grade II hydronephrosis**: Majority (almost 80–90%) can be managed conservatively. However, close monitoring of



Figs 39.24A and B: Anderson-Hynes reduction pyeloplasty (A) Excision of the redundant pelvis along with PUJ, (B) Completed ureteropelvic anastomosis

the patient is required to detect any deterioration in renal function. Any deterioration in renal function is an indication for surgical intervention. In this group, 10–20% patients benefit from early surgery (patients with renal function of involved kidney < 40%).

- **Grade III hydronephrosis**: All these patients should be operated early—Anderson-Hynes pyeloplasty to prevent permanent damage to kidney.

Types of pyeloplasty

1. Anderson-Hynes pyeloplasty

Principles

1. Excision of redundant pelvis
2. Disconnection of PUJ which is not functioning.
3. New ureteropelvic anastomosis is done in such a way that urine should drain by gravity.

- This is the most popular type of dismembered pyeloplasty (Figs 39.24A and B).

2. Nondismembered pyeloplasty

- They are Foley's Y-V plasty or flap pyeloplasty. The PUJ is not transected. However, they are not very popular.

RENAL TUBERCULOSIS

- This is secondary to pulmonary tuberculosis/lymphatic tuberculosis. The primary focus is often difficult to identify
  - Common in males, in the 20–40 age group.
  - Infection is always **haematogenous**. Often one may not find any active lesion in the lung or in the lymph nodes.
  - Usually unilateral.

Pathology (Figs 39.25 to 39.33)

1. Tubercles develop and coalesce over the papilla which may ulcerate—**ulcerative form**.
2. The tubercles may caseate and rupture over the renal papilla and communicate with the pelvis. This variety is called as **ulcerocavernous form**.
3. Attempt at healing produces calcification—**pseudocalculi** in the parenchyma of kidney.
4. Tubercular **hydronephrosis** is very rare. It is due to tubercular stricture of the pelviureteric junction.
5. The opening of one of the calyces may get fibrosed leading to **hydrocalyx** which may distort rest of the calyces.
6. Cortical abscess ruptures into the perinephric space and forms a tubercular **perinephric abscess.** This may even point at the loin and rupture forming a sinus in the loin.
7. Tubercular **pyonephrosis** (caseous kidney, putty kidney, cement kidney). When it gets calcified, it is called **cement kidney**. The entire kidney is converted into a bag of pus which is tubercular caseous material with or without secondary infection. The complete ureteric stricture due to tuberculosis cutting off the pelvicalyceal system may result in **autonephrectomy** because of fibrosis.
8. Small fibrosed, **contracted functionless kidney.**
9. As a part of **miliary tuberculosis**—multiple small tubercles may be seen in the renal parenchyma.

Clinical features

1. Frequency is the earliest symptom of tuberculosis. It is due to renal tubular inflammation and later due to tubercular cystitis.
2. Abacterial acid pyuria: The urine is opalescent, pale or yellow, acidic in reaction and no organisms/bacteria are grown on repeated culture.

PEARLS OF WISDOM

Sterile pyuria is seen in tuberculosis, stones and in carcinoma in situ.

3. Haematuria is not uncommon. Usually it is a small quantity due to ulcerocavernous variety.
4. Evening rise of temperature.
5. Loss of appetite and loss of weight.
6. Evidence of pulmonary or lymph node tuberculosis (TB) may be present.

Investigations

1. Urine for acid-fast bacilli (AFB)
   - Early morning sample of urine has to be examined which gives the highest concentration of AFB, for 3 days.
   - Ziehl-Neelsen stain and Gram staining
   - Lowenstein-Jensen media culture
   - Guinea pig inoculation is positive in 90% of cases.

PEARLS OF WISDOM

Newer easy methods like polymerised chain reaction (PCR) and radioisometric culture are also used.

A FEW PHOTOGRAPHS OF RENAL TUBERCULOSIS

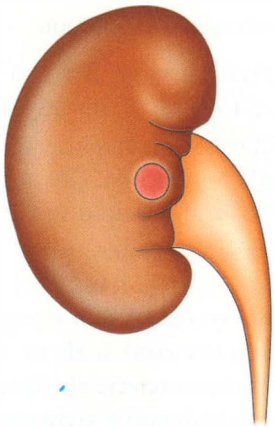


Fig. 39.25: Ulcerative form

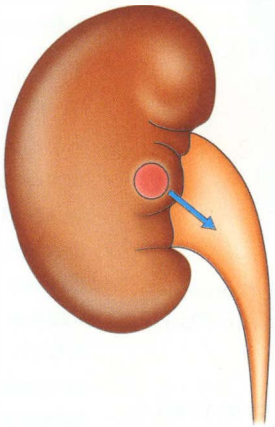


Fig. 39.26: Ulcerocavernous form

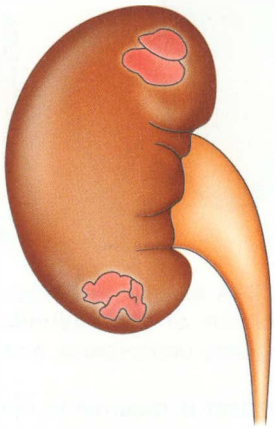


Fig. 39.27: Pseudocalculi

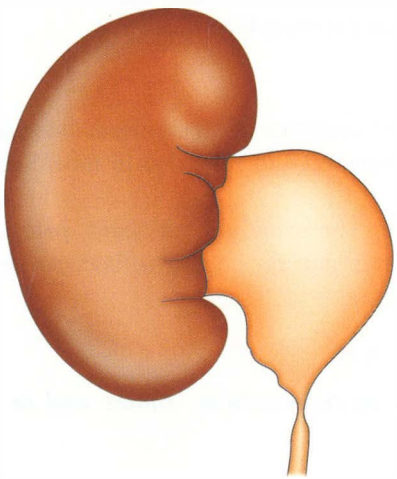


Fig. 39.28: Hydronephrosis

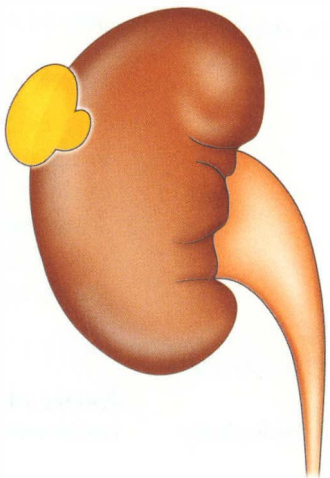


Fig. 39.29: Tuberculous perinephric abscess

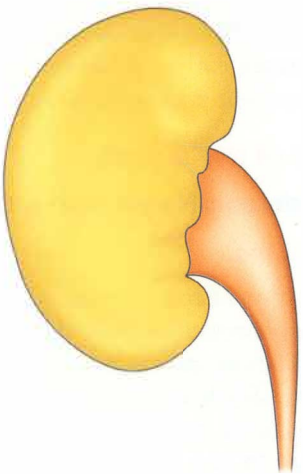


Fig. 39.30: Tuberculous pyonephrosis

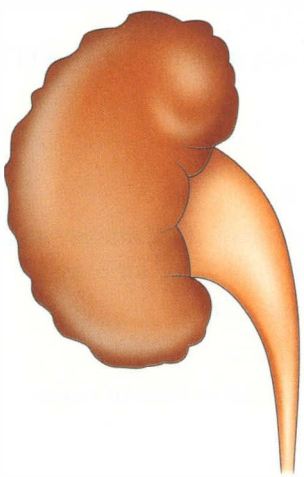


Fig. 39.31: Contracted kidney

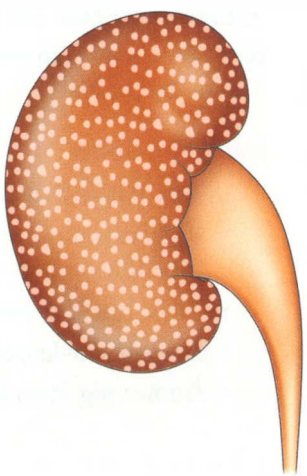


Fig. 39.32: Multiple tubercles—miliary tuberculosis

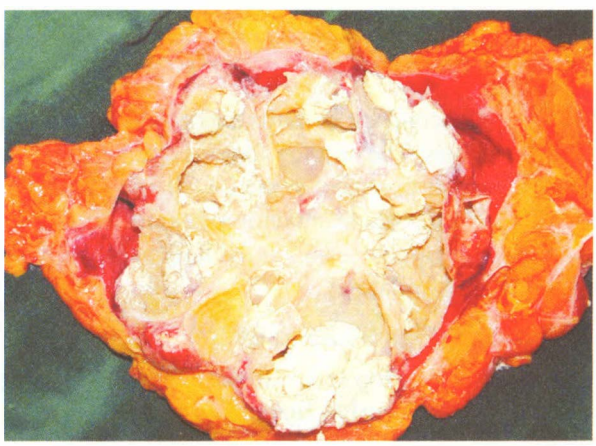


Fig. 39.33: Renal tuberculosis—entire kidney is a mass of caseous material

2. Cystoscopy

A. Initially small ulcers are seen around ureteric orifice. They join together resulting in a big ulcer.
   - Due to extensive periureteric fibrosis, the ureter becomes thickened, shortened and straight. The ureteric openings are lifted upwards and they are gaping which means it does not contract/does not close when bladder contracts. Such contracted, elevated, permanently opened, lower end of ureter is called as **golf hole ureter** (Fig. 39.34).
   - As a result of this, with each act of contraction of bladder, there is reflux of urine into the kidney causing damage,

B. When the disease involves the urinary bladder, it results in fibrosis. It becomes small and contracted with ineffective function. Storage capacity is lost resulting in intractable frequency, with a few drops of urine. There is bleeding. Micturition is painful and is called strangury. There is also severe pain in the suprapubic region which is referred to the tip of the penis. Such a small nonfunctioning urinary bladder is called as **thimble bladder** (Fig. 39.35).

Treatment

1. **Conservative line of management** with antituberculous treatment is successful, provided kidneys are functioning as in early stages.
2. **Nephroureterectomy** is indicated if the kidney is nonfunctioning (Fig. 39.36).
3. **Renal cavernotomy of Henley**
   - Indicated when there is stricture of calyces which results in a hydrocalyx.
   - In this operation the stricture is divided so that the drainage becomes better.
4. Treatment of **thimble bladder—ileocystoplasty**.
   - In this 10–15 cm of ileal loop is isolated based on the blood vessels, the fibrosed bladder dome is excised, intestine is split open and sutured to urinary bladder (Fig. 39.37).
   - This is to increase the capacity of bladder so as to store urine and reduce frequency.

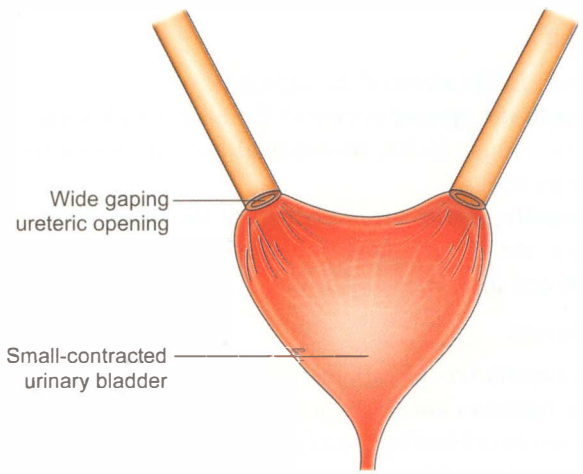


Fig. 39.34: Thimble bladder with golf hole ureter (TB)

Fig. 39.35: Genitourinary tuberculosis: Left kidney is normal. Right kidney shows hydroureteronephrosis. The urinary bladder is contracted—thimble bladder

RENAL NEOPLASMS

Classification

- **Benign:** Adenoma, cortical adenoma, papilloma arising from pelvis, haemangioma.
- **Malignant:** Nephroblastoma, renal cell carcinoma
- Transitional cell carcinoma (rare)
- Squamous cell carcinoma (extremely rare)

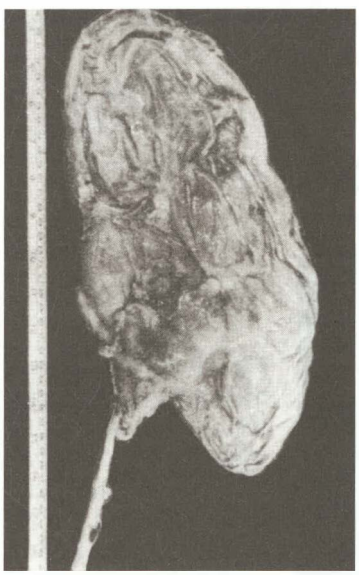


Fig. 39.36: Nephroureterectomy for renal tuberculosis (*Courtesy:* Professor Sasidharan, Manipal)

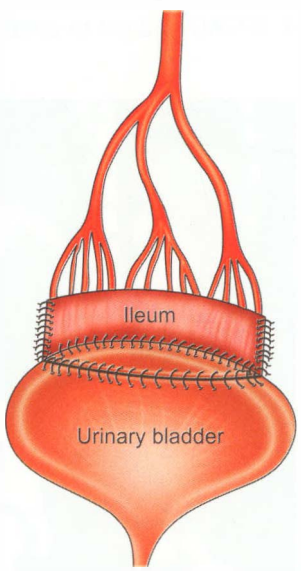


Fig. 39.37: Ileocystoplasty

WILMS' TUMOUR (NEPHROBLASTOMA)

- This is a malignant tumour of the kidney occurring in **children**.
- The tumour is composed of epithelial and mesothelial elements. Thus, it may contain bone, cartilage, muscle, etc. Hence, it is called **nephroblastoma (immature embryonic tissue)**.
- The tumour arises in one of the poles, distorting the reniform shape of the kidney. It is greyish white or pinkish white in colour. At places, there may be areas of haemorrhage/necrosis.
- Microscopic features include connective tissue elements cartilage, spindle cells, smooth striated muscle cells and epithelial elements.

Clinical features

- Common in **female children**, around 2–4 years.
  - Less than 1 year of age carries good prognosis
  - Upper limit of age is 7 years
  - Rarely it may occur in adolescents
- The child is brought with **abdominal distension**, due to hugely enlarged kidney which on palpation feels nodular.
- Rarely, Wilms' tumour can be bilateral
- **Haematuria** is a bad prognostic symptom. It is an indication of rupture of tumour into the pelvis of kidney. Most of such children die by 2 years of age.
- **Low grade fever** can occur in rapidly growing tumour due to tumour necrosis, which releases pyrogens.
- Rapid deterioration of health is characteristic.

Investigations

1. Abdominal USG can detect a solid tumour in the kidney. Ultrasound also rules out opposite kidney tumour.
2. CT scan to know extent of lesion and spread to the adjoining structures (Fig. 39.38).
3. IVP is done to study distortion of calyces and to evaluate the function of the opposite kidney.
4. FNAC is done to confirm the diagnosis preoperatively.

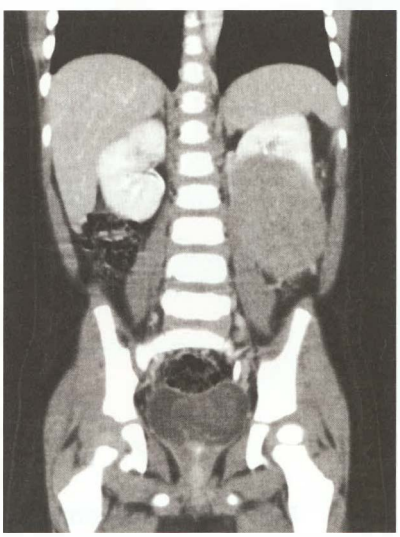


Fig. 39.38: A 3-year-old male child had presented with left loin mass. CECT abdomen and pelvis was done which showed a large tumour in the left kidney. Left radical nephrectomy was done and the histopathology was reported as Wilms' tumour

Differential diagnosis

1. **Neuroblastoma** arises from adrenals. This is more common than nephroblastoma.
2. **Retroperitoneal tumours**
3. Adrenal tumour (Fig. 39.39)

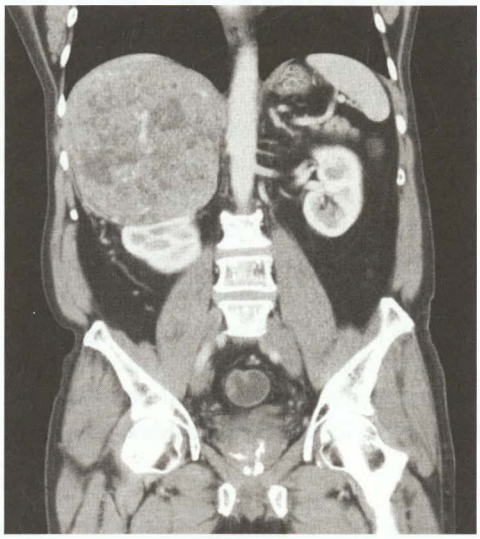


Fig. 39.39: CECT abdomen and pelvis showing a large right heterodense adrenal tumour

Differentiating features between Wilms' tumour and neuroblastoma (NBL)

1. **Calcification**—foci of calcification seen in NBL (85%). Less common in Wilms' (15%).
2. **Intraspinal extension**—seen in NBL
3. **Aorta and IVC** invasion by Wilms'
4. **Location**
   Wilms'—intrarenal
   Neuroblastoma—seen above kidney pushing it downwards and outwards.
5. Crossing midline—neuroblastoma
6. Homovanillic acid (HVA) and Vanillylmandelic acid (VMA) increase in neuroblastoma.

Spread

1. **Direct infiltration** of the capsule
2. **Lymphatic spread** occurs to the hilar lymph nodes, para-aortic lymph nodes, mediastinal and left supraclavicular lymph nodes.
3. **Haematogenous spread** occurs to the lungs, liver, bones, brain, etc. The tumour thrombus can extend to the renal vein and inferior vena cava.

Treatment

The anaemia has to be corrected at the earliest.

1. **For tumours confined to renal capsule** or perirenal soft tissue not infiltrating the adjacent organs, **radical nephrectomy** followed by chemotherapy with actinomycin D and vincristine are given for 6 months.

2. **For tumours which have gone beyond renal capsule** and perirenal soft tissue, local infiltration to adjacent tissue, lymphatic metastasis, nephrectomy followed by local radiotherapy and chemotherapy is given with actinomycin D and vincristine for 15 months.
3. **If the tumour is found to be unresectable** by CT scan or magnetic resonance imaging (MRI), preoperative FNAC to confirm diagnosis is indicated followed by preoperative radiotherapy (1000 cGy) or chemotherapy. Once the tumour regresses in size, nephrectomy has to be done. Postoperative chemotherapy is given with actinomycin D, vincristine and doxorubicin.
4. **Bilateral Wilms' tumour:** Radical nephrectomy on the side of the larger tumour and partial nephrectomy on side of the smaller tumour should be done. As much of renal tissue as possible should be preserved after leaving a tumour free margin. Postoperatively the patient has to be treated with chemotherapy. If the surgery is not feasible, only radiotherapy and chemotherapy has to be given. Growth disturbances, cardiac and pulmonary toxicities are complications of radiotherapy.

RENAL CELL CARCINOMA (RCC)

- It is also called **Hypernephroma** or **Grawitz tumour.**
- Commonly found in the age group 40–60.
- Male : female ratio 2 : 1

AETIOLOGY (Key Box 39.6)

- Not known. Rarely **familial RCC** is known. Specific oncogenes have been identified. Smokers are twice as often affected than nonsmokers.
- **Leather workers** are more prone for RCC.
- **Analgesic abusers** are also more prone.

KEY BOX 39.5

RISK FACTORS OF RCC

- Diabetes mellitus
- Chronic dialysis

KEY BOX 39.6

RENAL CELL CARCINOMA: AETIOLOGY

1. **C**hronic cystic disease
2. **C**hromosomal defect
3. **C**admium exposure
4. **C**igarette smoking
5. **C**offee drinking
6. **C**ongenital disease—von Hippel-Lindau disease

Observe **6 Cs**

PATHOLOGY

- Nearly all renal cancer in adults are **adenocarcinoma**
- Cell of origin: **Proximal renal tubular epithelium**.
- Starts in one of the poles, **commonly in the upper pole** and usually ruptures outside the capsule because of which **reniform shape of kidney is maintained** (Wilms' tumour grows within the capsule. Hence, the kidney shape is lost very early) (Fig. 39.40A).
- On the outer surface, it is homogenous (Wilms' tumour is pleomorphic) and yellow in colour due to position of lipids.
- A few **haemorrhagic areas** are common because the tumour is very vascular (Fig. 39.40B).
- Microscopy: **Alternate clear cells and dark cells.**
- **Various subtypes of RCC**
  1. **Clear cell carcinoma**
     - Most common type (70–80%)
     - It can be familial, associated with von Hippel-Lindau syndrome or sporadic (95%).
  2. **Papillary**—both familial and sporadic forms.

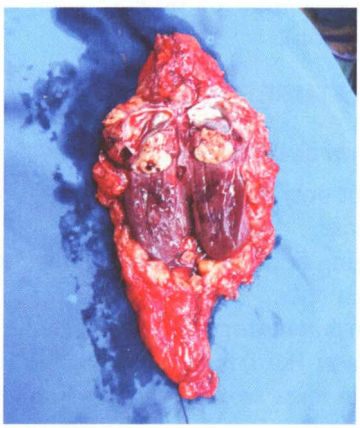


Fig. 39.40A: Renal cell carcinoma upper pole

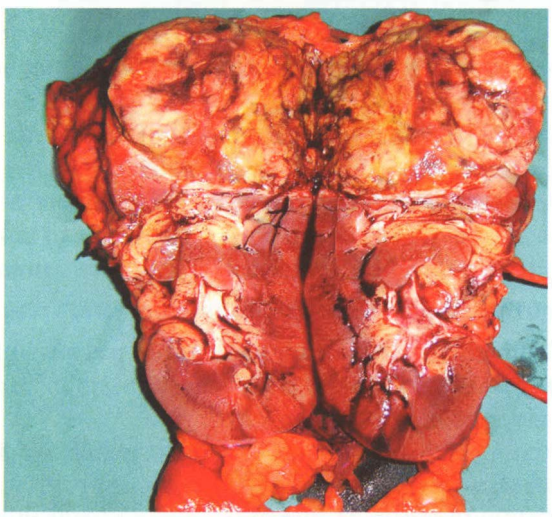


Fig. 39.40B: Upper pole RCC—*see* the fleshy tumour with haemorrhage

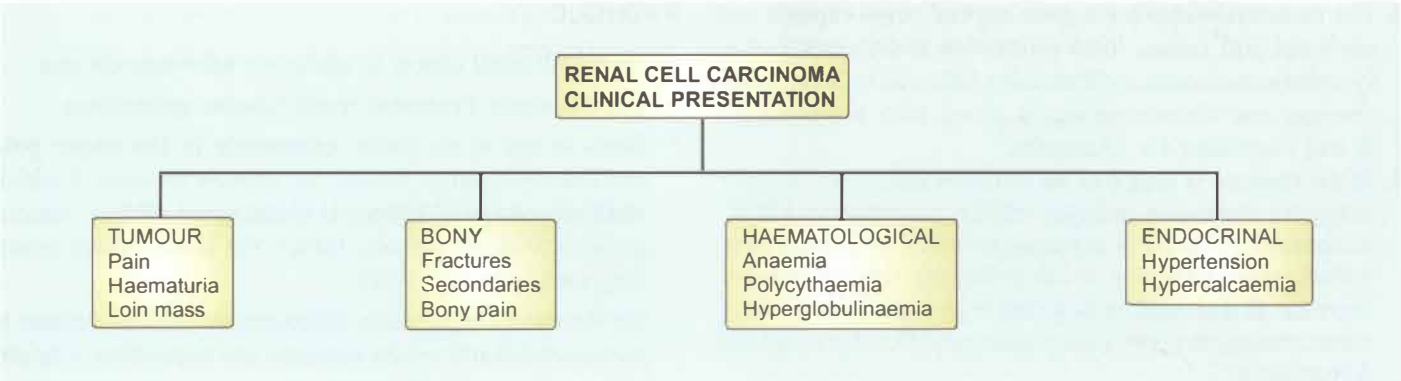


Fig. 39.41: RCC showing clinical presentation

3. **Chromophobe**—these tumours exhibit multiple chromosome losses and extreme hypoploidy.
4. **Renal medullary carcinoma**
   - new subtype
   - associated with sickle cell trait

- ***Tumour cells line the blood vessels*** which are responsible for early blood spread from renal cell carcinoma (like follicular carcinoma of thyroid—angioinvasion and capsular invasion).

CLINICAL FEATURES (Fig. 39.41)

I. Triad of renal cell carcinoma

Triad is seen in only 9%, but if present, strongly indicates metastatic disease.

1. **Pain:** Dragging or intermittent clot colic due to blood clot blocking the ureter.
2. **Intermittent haematuria**
3. **Palpable mass:** Hard, nodular, ballotable and bimanually palpable, loin mass moving with respiration.

II. Other manifestations

1. **Pathological fractures**, e.g. fracture femur, humerus vascular, pulsatile, secondaries are common in the flat bones, e.g. scalp, vertebra, rib, sternum because they contain red marrow for longer time.
2. **Anaemia** disproportionate to amount of haematuria is due to decreased production of erythropoietin.
3. Mild elevation of **temperature** is due to tumour necrosis producing pyrogens. It can present as pyrexia of unknown origin (PUO) and hence it is called **Internist's tumour.**
4. **Nephrotic syndrome**—like features are rare.
5. **Endocrinal disturbances** are rare (**paraneoplastic syndromes**). Increased ESR—most common paraneoplastic syndrome.
   - Renin producing tumours are responsible for **hypertension**.
   - **Polycythaemia** is due to increased erythropoietin secretion.
   - Other hormones produced by the tumour are parathormone, adrenocorticotrophic hormone, human chorionic gonadotropin, glucagon, prolactin.
6. **Hypertension**
7. **Liver dysfunction:** Nonmetastatic liver dysfunction also known as **Stauffer's syndrome and this improves after nephrectomy.**

Robson's staging of renal cell carcinoma

- Stage I: Tumour limited to kidney
- Stage II: Tumour invades perinephric tissues or adrenal gland, but does not extend beyond Gerota's fascia.

TNM STAGING RENAL CELL CARCINOMA

Tx	Primary tumour cannot be assessed
T0	No tumour
T1	< 7 cm, within capsule, in greatest dimension
T2	> 7 cm, within capsule
T3a	Extracapsular but within Gerota's fascia extension
T3b	Into adrenal/perinephric
T3c	Extension into renal vein/IVC
T4	Extension above diaphragm—direct invasion beyond Gerota's fasica
N0	No nodes
NX	Cannot be assessed
N1	Single regional lymph node
N2	More than one regional lymph node
M0	No metastasis
M1	Distant metastasis present

Stage grouping

Stage I	T1 N0 M0
Stage II	T2 N0 M0
Stage III	T1 N1 M0
	T2 N1 M0
	T3 N1 M0
Stage IV	T4 any N, M0
	Any T, N2, N3, M0
	Any T, any N, M1

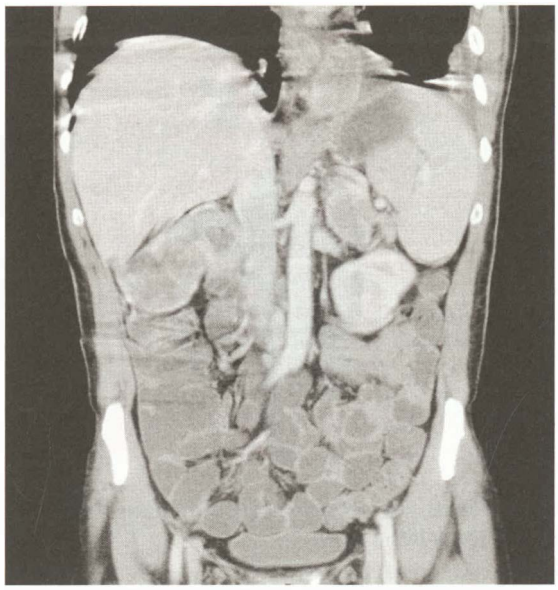


Fig. 39.42: Right RCC with left adrenal metastasis

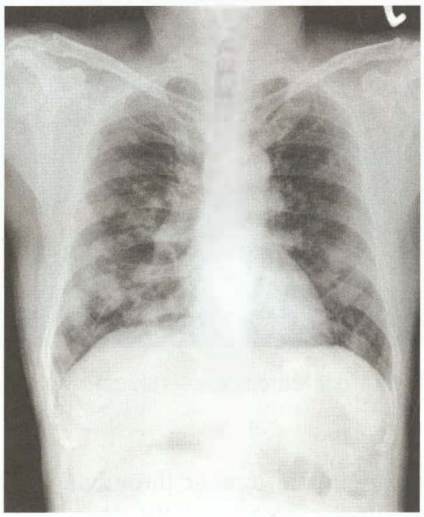


Fig. 39.43: Chest X-ray showing cannon ball

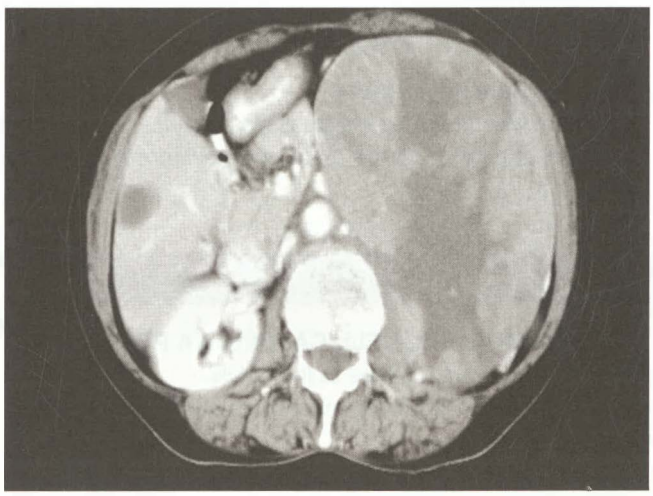


Fig. 39.44: CT scan showing left RCC

- Stage III : Tumour extends into major veins or lymph noda involvement
- Stage IV: Tumour invades beyond Gerota's fascia or distan metastasis.

INVESTIGATIONS

1. **Urine examination** is done when the patient has haematuria to look for malignant cells.
2. **Plain X-ray KUB region:** Enlarged kidney can be seen.
3. **IVP:** Distortion of calyces, missing of calyces or loss of architectural pattern of kidney.
4. **USG:**
   - Enlarged kidney
   - Locate tumour, site and extent
   - USG-guided FNAC can be done
   - Can detect thrombus in inferior vena cava (IVC)
5. **Contrast enhanced CT scan is the investigation of choice** for staging (Key Box 39.7 and Figs 39.42 to 39.46).

KEY BOX 39.7

CT SCAN

- Mixed density mass lesion
- Enhancement after contrast
- Secondary changes such as tumour cell necrosis
- Local extent can be evaluated
- IVC thrombus and lymph node involvement identified.

PEARLS OF WISDOM

Any renal mass which enhances after contrast on a CT scan in an elderly person should be considered as RCC until proved otherwise.

6. **Renal angiography** is done by retrograde transfemoral approach. Features are as follows:
   - Neovascularisation: Tumour blush inside the tumour.
   - Venous phase has to be observed to rule out tumour extension in the vein.

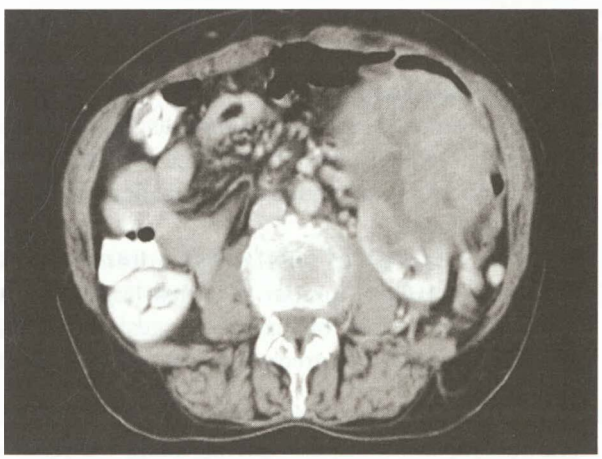


Fig. 39.45: Very extensive left RCC

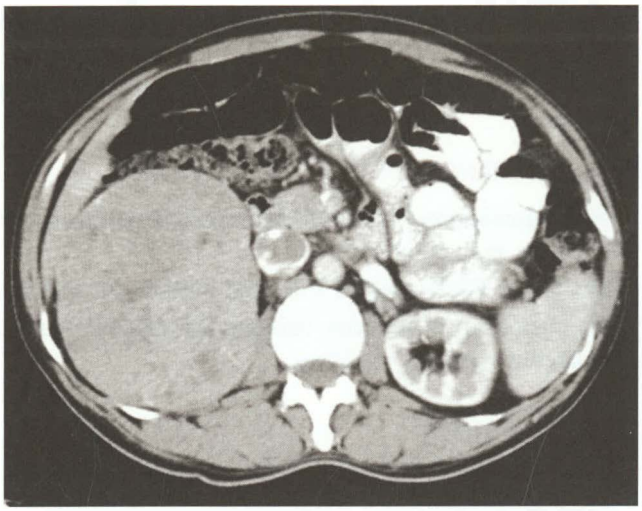


Fig. 39.46: IVC thrombus

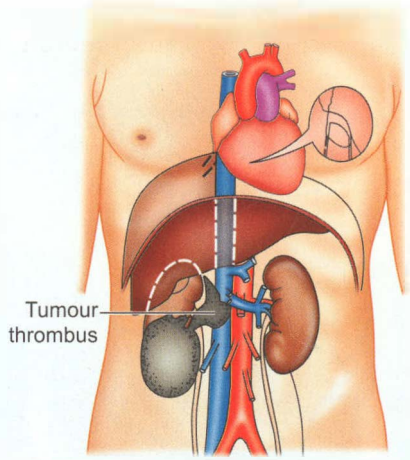


Fig. 39.47: Renal cell carcinoma with tumour thrombus in the renal vein, inferior vena cava

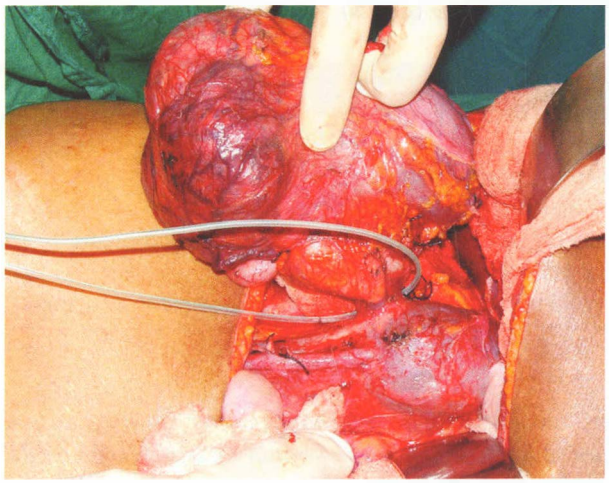


Fig. 39.48: Renal cell carcinoma with tumour thrombus removed

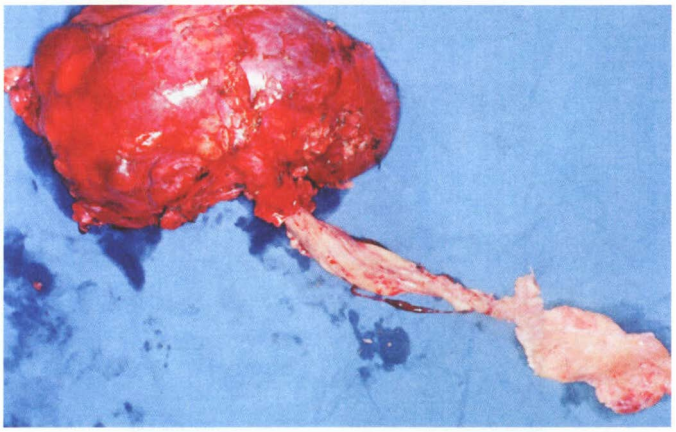


Fig. 39.49: Renal cell carcinoma—radical nephroureterectomy specimen

7. MRI scan: MRI scan is the investigation of choice to know the extent of IVC thrombosis (better than CT).

8. Venacavogram: It is done to know extent of tumour in the IVC and presence of collateral circulation.

TREATMENT

1. Radical nephrectomy

- *En bloc* removal of entire Gerota's fascia with its contents, i.e. kidney, proximal ureter, **adrenal gland**.
- Retroperitoneal lymph node dissection does not improve the survival rate.
- Routine removal of ipsilateral adrenal gland is uncommon. Unless tumour invovles large portion of upper pole of kidney or there is suggestion of adrenal gland involvement on preoperative radiologic exams.

2. Radical nephrectomy with extraction of tumour thrombus

- The tumour thrombus can extend along the renal vein into IVC and even into the right atrium (Fig. 39.47).
- Infradiaphragmatic tumour thrombus can be removed with proximal control over the vena cava. The supra-diaphragmatic IVC thrombus requires cardiopulmonary bypass (Figs 39.48 and 39.49).
- In the absence of distant metastasis after removal of the thrombus these patients survive for a long duration.

3. Nephron sparing surgery

It is done when tumour length is less than 5 cm, in bilateral tumours and in cases of renal cell carcinoma in solitary kidney.

KEY BOX 39.6

TRANSITIONAL CELL CARCINOMA PELVIS

- Uncommon tumour arises in the pelvis
- Multiple sites of urothelial mucosa involvement often.
- Low grade tumours
- Discovered late
- Haematogenous spread is common.

4. Therapeutic embolisation

- This can be used as a palliative measure in advanced carcinoma to relieve symptoms. This can also be used preoperatively to regress the size of the large tumour.
- A catheter is placed in the renal artery and substances such as gel foam, blood clot, crushed muscle are injected.
- They block the lumen of the vessel and reduce size of the tumour so that radical nephrectomy can be undertaken later.

5. Radiotherapy

Not of much use. However, it is a good form of palliation for secondaries in the lung, bone and brain.

6. Immunotherapy

Administration of **interferon** or **interleukin-2** has been found to **improve the survival rate.**

RENAL MASS IN SURGICAL WARD (Table 39.2)

Clinical features of kidney mass

- Moves with respiration because the fascia of Gerota encloses the kidney and fuses above with the diaphragm.
- Kidneys enlarge in the upward and downward direction.
- Bimanually palpable and ballotable. It is ballotable because of renal pedicle and perirenal pad of fat.
- Colonic band of resonance is obliterated when the kidney enlarges, as the colon is pushed laterally.
- Upper border is not palpable because it is under the 12th rib.

ACUTE SURGICAL INFECTIONS OF THE KIDNEYS

PYONEPHROSIS

In this condition, the entire kidney is converted into a sac containing pus or purulent urine—almost always the renal parenchyma is destroyed totally.

Causes

1. **Renal calculous disease** is the most common cause of pyonephrosis.
2. **Acute pyelonephritis** is more common in children and in females. Inadequately treated cases may develop into pyonephrosis, especially when pyelonephritis is associated with urinary tract obstruction.
3. **Infection** of a hydronephrosis

Clinical features

- Anaemia and fever
- Renal swelling
- Large swelling with high grade fever with chills and rigors suggest an imminent danger of septicaemia and calls for an immediate drainage of the pus.

Investigations

- Urine examination may be positive for coliforms and other gram-negative organisms.
- Plain X-ray KUB may reveal a stone or an enlarged renal outline.
- Ultrasound can confirm hydronephrosis.

Table 39.2 Renal mass in surgical ward

	Hypernephroma	Hydronephrosis	Polycystic kidney
1. Chief symptoms	Haematuria, pain in the loin, renal mass	Asymptomatic, distension, abdomen pain	Mass abdomen, hypertension, haematuria
2. Age of the patient	Over 50 years	20–30 years	30–40 years
3. Sex incidence	Common in males	Common in females	Common in females
4. Anaemia	Present	Absent	May be present
5. Features of renal failure	Absent	Can be present in bilateral cases (rare)	May be present
6. Renal mass	Unilateral, nodular, hard, may be fixed, nontender	Can be bilateral, smooth, cystic, feels firm	Bilateral, bosselated, nodular, not fixed, nontender
7. Features of kidney mass	May not have free mobility due to fixity	Not fixed, nontender	Present
8. IVU	Irregular calyces	Gross dilatation of pelvicalyceal system	Spider-leg deformity of calyces
9. CT Scan	Enhancing mass	Dilated pelvicalyceal system with uniform filling of contrast	Multiple hypodense areas without enhancement
10. Treatment	Radical nephrectomy	Pyeloplasty	Symptomatic—renal transplantation

- Intravenous urogram demonstrates poor function of the kidney on the diseased side. As a rule the opposite kidney is normal.

Treatment

- Broad-spectrum antibiotics (parenteral) should be started immediately once the urine and blood is sent for culture and sensitivity.
- Ultrasound-guided aspiration of pus or a percutaneous nephrostomy (better), and drainage of pus greatly improves the general condition of the patient.
- If any obstruction or causative agent such as a stone is found, it should be removed.
- Nephrectomy should be considered if the kidney is non-functioning with significant damage.

PERINEPHRIC ABSCESS

It refers to the collection of pus in the perirenal area.

Causes

- Infection in a perirenal haematoma
- Pyonephrosis when it ruptures
- Tubercular perinephric abscess
- Pus from retrocaecal appendicitis can extend into loin, perinephric area and may present as abscess.

Clinical features

- High swinging temperature
- Rigidity, tenderness, fullness in the loin
- Oedema in the loin

Investigations

- Total count: Raised above 20,000 cells/mm^3
- Urine analysis: No organisms are usually found
- X-ray spine: Scoliosis with concavity towards abscess
- Screening chest: Diaphragm is immobile and elevated on the diseased side.

Treatment

- Pus is drained by an incision in the loin, breaking all the loculi.
- Dialysis and renal transplantation are discussed in page 1111.

MISCELLANEOUS

INTERESTING 'MOST COMMON' FOR RENAL CELL CARCINOMA

- Most common renal cancer in adults are adenocarcinomas.
- Most common site is upper pole of the kidney.
- Most common presentation of renal cell carcinoma is with mass abdomen.
- Most common investigation of choice is contrast enhanced CT scan.
- Most common method of spread is by haematogenous route.
- Most common intra-abdominal malignancy which spreads within vena cava and into atrium is renal cell carcinoma.
- Most common cell of origin is proximal renal tubular epithelium.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been updated.
- New photographs and key boxes have been added.

MULTIPLE CHOICE QUESTIONS

1. Following is not the feature of adult polycystic kidney:

A. It can give rise to renal failure

B. Hypertension is seen in about 75% patients

C. It is autosomal recessive

D. It is always bilateral

2. Relationships of right kidney include following *except*:

A. Posteriorly are muscles

B. Anteriorly pyloric antrum

C. Lateral ascending colon

D. Medial adrenals

3. Which of the following is the feature of horseshoe kidney?

A. Classically it is the upper polar fusion of both kidney

B. It does not cause angulation of the ureter causing hydronephrosis

C. It can be associated with Down's syndrome

D. Hyperextension of the spine results in pain, nausea and vomiting

4. Gout results in:

A. Calcium stones B. Cystine calculi

C. Phosophate stones D. Uric acid stones

5. Which type of stone typically causes haematuria?
A. Oxalate stone B. Phosphate stone
C. Cystine stone D. Uric acid stone

6. Following are true for calcium oxalate stone *except*:
A. They are called mulberry calculi
B. It is smooth and round
C. It is hard
D. It is visible in X-ray

7. About uric acid stone following is true *except*:
A. It is common in those who consume red meat
B. Small and multiple
C. Also seen in gout
D. Pure uric acid stone is radio-opaque

8. Staghorn calculi is a:
A. Calcium oxalate stone
B. Phosphate stone
C. Uric acid stone
D. Cystine calculus

9. Following are true for upper ureteric stone *except*:
A. ESWL is the ideal treatment
B. Percutaneous removal is an ideal treatment
C. Ureteroscopic removal is an ideal treatment
D. Cystoscopic removal is ideal

10. Patient has renal stones and swelling in the neck, the diagnosis can be:
A. Medullary carcinoma thyroid
B. Hyperparathyroidism
C. Hyperthyroidism
D. von Recklinghausen's disease

11. Which of the following is the cause for bilateral hydronephrosis?
A. Idiopathic retroperitoneal fibrosis
B. Carcinoma ureter
C. Tuberculosis of urinary tract
D. Aberrant artery

12. Following are features of Dietle's crisis *except*:
A. It is called intermittent hydronephrosis
B. It is usually seen in calculus hydronephrosis
C. Does not produce a mass
D. It happens due to slippage of stone

13. Which is the investigation of choice in hydronephrosis?
A. Ultrasound B. Intravenous pyelography
C. CT scan D. Isotope renography

14. Definite indication of pyeloplasty include following *except*:
A. Calculus hydronephrosis
B. Grade III hydronephrosis
C. Nonfunctioning kidney with cortical thickness 0.1 cm
D. Tuberculous hydronephrosis

15. Following are true for renal tuberculosis *except*:
A. It is primary tuberculosis
B. Infection occur through lymphatic spread
C. Commonly causes hydronephrosis
D. It causes bacterial acid pyuria

16. Which of the following is an uncommon symptom of renal tuberculosis?
A. Haematuria
B. Evening rise of temperature
C. Cystitis
D. Pyuria

17. Following are the result of urinary tuberculosis *except*:
A. Thimble bladder B. Golf hole ureter
C. Putty kidney D. Hydronephrosis

18. Following are true for Wilms' tumour *except*:
A. Tumour contain only epithelial elements
B. Common age group is less than 1 year of age
C. Prognosis is poor if it occurs within 1 year
D. Haematuria is a bad prognostic sign

19. About renal cell carcinoma all are true *except*:
A. Majority in adults are adenocarcinoma
B. Reniform shape is maintained
C. Clear cells and dark cells are present in microscopy
D. Tumour cells do not line the blood vessels

20. Polycythaemia in renal cell carcinoma is due to:
A. Renin production
B. Erythropoietin production
C. Prolactin production
D. Glucagon production

21. The diagnostic CT finding of a renal cell carcinoma in an elderly patient is:
A. Dense mass lesion
B. Tumour necrosis
C. Lymph nodes
D. Enhancing mass after intravenous contrast

22. Following are true for transitional cell carcinoma *except*:
A. Low-grade tumours
B. Haematogenous spread is common
C. Arise from urothelium
D. Multiple sites are not commonly involved

ANSWERS

1 C	2 B	3 D	4 D	5 A	6 B	7 D	8 B	9 A	10 B
11 A	12 C	13 C	14 B	15 D	16 A	17 D	18 D	19 D	20 B
21 D	22 D								

40

The Urinary Bladder and Urethra

- Surgical anatomy
- Vesical calculus
- Carcinoma of bladder
- Ectopia vesicae
- Acute cystitis
- Diverticula
- Urinary fistulae
- Interstitial cystitis
- Schistosoma haematobium
- Urinary diversion
- Rupture bladder
- Surgical anatomy of urethra
- Rupture urethra
- Stricture urethra
- Hypospadias
- Retention of urine
- Posterior urethral valve
- What is new?/Recent advances

SURGICAL ANATOMY OF THE BLADDER

Lining epithelium

- Urinary bladder is lined by transitional epithelium that covers a connective tissue known as lamina propria.
- **Bladder cancers are transitional cell carcinomas.**
- However, due to changes in the epithelium caused by chronic irritation (stone), one can get other malignancies such as squamous cell carcinoma.
- Bladder cancer can easily spread through lamina propria into the muscle coat (**detrusor muscle**).

Detrusor muscle

It is a smooth muscle, fibres of which are intermingled. Hence, in cases of bladder neck obstruction, changes such as trabeculations/sacculations due to hypertrophy of this muscle is found.

Trigone

It is a triangular area lying between the internal urethral orifice and the orifices of the ureter. It is the **most sensitive part of the bladder,** irritation of which is mainly responsible for frequency of micturition (Fig. 40.1).

Bladder neck

- Internal sphincter is the smooth muscle which surrounds the bladder neck. It is innervated by α-adrenergic fibres and prevents retrograde ejaculation.
- Distal urethral sphincter is a somatic striated muscle which is supplied by S2–S4 fibres *via* pudendal nerves.

Supports of the bladder

- Posteriorly **endopelvic fascia,** which is continuous with lateral ligaments of the rectum, need to be divided during radical cystectomy.
- Anteriorly **puboprostatic ligaments** also need to be divided during radical cystectomy.

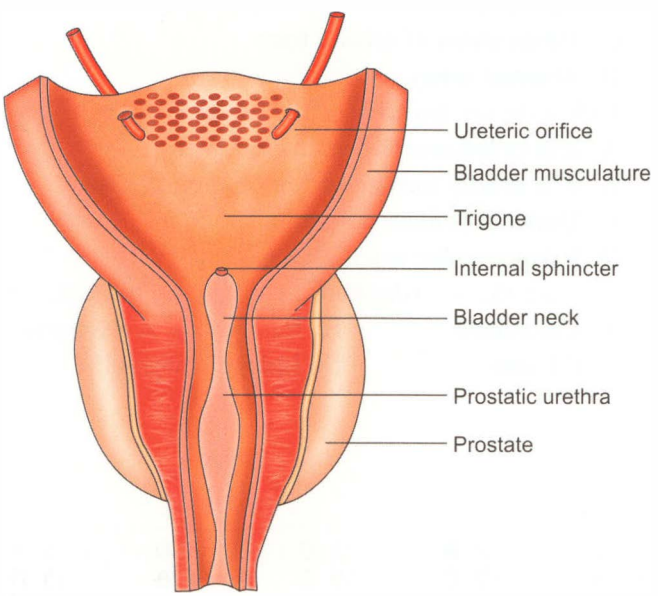


Fig. 40.1: Surgical anatomy of the urinary bladder

- **False ligaments:** Urachus and obliterated hypogastric arteries together with the fold of peritoneum overlying these structures are called as false ligaments (median and lateral ligaments).

Blood supply

- **Superior and inferior vesical arteries** are derived from anterior trunk of internal iliac artery as the main source of arterial blood supply. Minor blood supply comes from obturator, inferior gluteal and in females from uterine and vaginal arteries also.
- **Veins** form plexus on lateral and inferior surfaces of the bladder. Hence during suprapubic cystostomy, these structures have to be avoided while entering into the bladder.
- **Vesical plexus** is continuous with prostatic plexus of veins in males which drain into the internal iliac vein.

Lymphatics

- Internal iliac nodes are the first level of lymph nodes.
- Obturator and external iliac lymph nodes get involved later.

Innervation

- **Parasympathetic** comes from anterior divisions of sacral nerves—S2,S3,S4 through inferior hypogastric plexus. Following excision of rectum, disturbance of micturition and sexual function can occur due to damage to the pelvic plexus.
- **Sympathetic** supply comes from T10 to L2 segments.

VESICAL CALCULUS

- **Primary:** Stone which develops in sterile urine. They develop in the absence of bladder pathology. These also include renal stones which have migrated to the bladder.
- **Secondary:** Stone develops in the presence of infection and stasis due to obstruction to the urinary flow. They develop secondary to bladder pathology.

Types (Figs 40.2 and 40.3)

1. **Oxalate stone:** Moderate size, uneven surface, mulberry stone is dark brown or black because of incorporation of blood pigment in it.
2. **Uric acid stone:** Round to oval, smooth, pale yellow, not opaque to X-rays. They are primary stones.
3. **Cystine:** Radio-opaque due to high sulphur content.
4. **Triple phosphate:** These stones consist of ammonium, magnesium and calcium phosphates. **They occur in urine infected with urea-splitting organisms**. Sometimes, they grow rapidly. The nucleus of the stone can be made up of bacteria, desquamated epithelium or a foreign body. Dirty white in colour.

Clinical features

- **Frequency** of micturition is the earliest symptom of bladder stone. It is due to cystitis.
- **Pain** at the end of micturition referred to tip of penis in young boys suggests bladder stone. In school-going children, pain is aggravated by jumping and jolting. Pain is decreased on lying down because stone falls away from the trigone of the bladder. Typically, oxalate stones produce pain. Painful ineffective micturition is described as strangury.
- **Haematuria,** is due to stone causing abrasions in the bladder mucosa.
- **Acute retention of urine** due to the calculus obstructing the internal meatus.
- **Males** are affected 8 times more frequently than females.

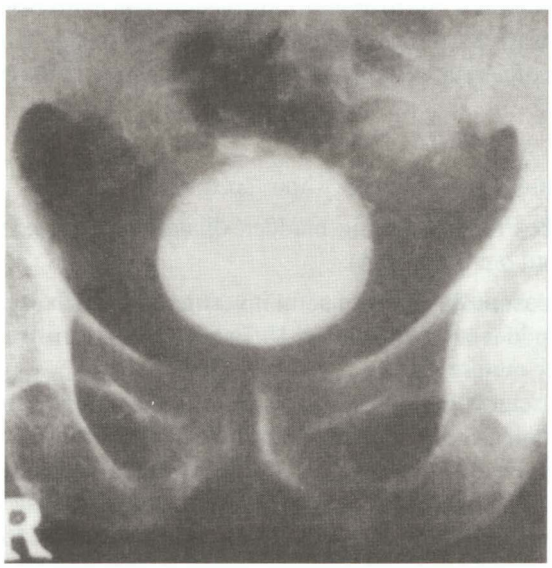


Fig. 40.2: Bladder stone—triple phosphate stone

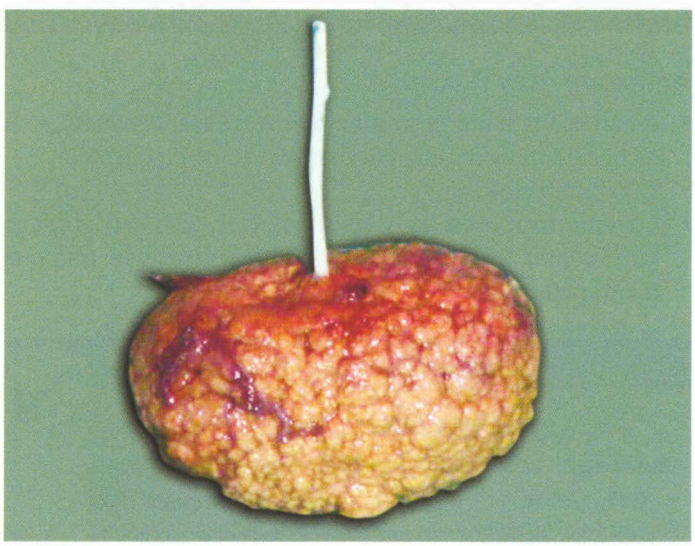


Fig. 40.3: Bladder stone with copper-T (*see* clinical notes in page 970)

Investigations

- **Urine:** Red blood cells may be present—microscopic haematuria
  - Envelope-like crystals: Oxalate stone
  - Hexagonal plates: Cystine calculi
- **Radiography:** In 90% of cases, the stone is visible. However, it is important to look for stones in the entire urinary tract.
- **Cystoscopy:** Stone can be visualised.
  A **click** can be heard when the stone comes into contact with the instrument.

Treatment

- Ultrasound lithotripsy — very safe, but only for small stones.
- Laser lithotripsy (Holmium laser)—can break most large stones.
- Percutaneous suprapubic litholapaxy—using needle, guidance and metal dilators.

I. Litholapaxy

- By introducing a cystoscopic lithotrite, stone is grasped firmly and broken. Small fragments of stone are evacuated by using evacuator.

Contraindications for litholapaxy

1. **Urethra:** Obstruction such as stricture, enlarged prostate.
2. **Bladder:** Cystitis, contracted bladder, carcinoma.

II. Suprapubic cystolithotomy

- Can be done when the stone is too big, too hard to crush or too soft.
  Transitional cell Ca — 90%
  Squamous cell Ca — 5–10%
  Adenocarcinoma — 2%

CARCINOMA OF THE BLADDER

Aetiology (Key Box 40.1)

- Incidence is more in **aniline dye workers**. Products such as benzidine and 3-naphthylamine are carcinogenic.
- Cigarette **smoking**
- Chronic **irritation** by stones, catheter, also can produce carcinoma: 95% of the tumours originate in mucous membrane.

KEY BOX 40.1

BLADDER CANCER

Aetiology	*High risk—occupations*
• Bilharziasis	• Aniline dye workers
• Chronic irritation	• Leather industry workers
• Chronic smoking	• Paint industry workers
• Cyclophosphamide	• Rubber industry workers

- **Bilharziasis or schistosomiasis** increases the chances of bladder cancer (squamous cell carcinoma).
- **Congenital anomalies** associated with increased risk of carcinoma bladder
  a. Patent urachus
  b. Exostrophy bladder

Pathology

1. **Malignant villous tumours:** They are transitional cell carcinomas. Multiple primaries are found in 25% of patients with bladder cancer.
   a. The villi are stunted, swollen and resemble cauliflower. They are slow growing.
   b. Can be sessile. Such tumours are high grade.
   c. Bladder wall is more vascular.
   d. Submucous lymphatic nodules appear around the growth.
2. **Solid tumours are always malignant:** They are sessile, lobulated.
3. **Carcinomatous ulcer:** It arises in **leukoplakia**.

Histological types

1. Transitional cell carcinoma—90% of tumours
2. Squamous cell carcinoma
3. Adenocarcinoma arises from urachal remnants and urethral glands
4. Mixed variety
5. Undifferentiated.

Clinical features

- In 90% of cases, initial symptom is **painless, intermittent haematuria**.
- **Severe cystitis** like symptoms occur in carcinomatous ulcer.
- Later painful, **blood-stained micturition** can occur.
- **Strangury:** Painful micturition with bleeding and incomplete emptying of bladder.
- **Loin pain** is due to ureteric obstruction with hydronephrosis.
- **Suprapubic pain,** groin pain, perineal pain are due to infiltration of nerves. This indicates advanced nature of the growth.

Investigations

1. **Urine:** Cytology of a 3-hour specimen.
2. **IVP:** Filling defect in bladder, dilatation of ureter can be found out (Fig. 40.4).
3. **Ultrasound** is a very useful investigation which can detect a bladder carcinoma (Figs 40.7 and 40.8). It can also detect liver metastasis.
4. CT scan is the investigation of choice especially to know the spread of the disease (Figs 40.5, 40.6 and 40.9 to 40.11).
   - Specially it is useful to know the infiltration of the muscle, perivesical tissue and also the prostate and pelvic wall.
5. **Cystoscopy: It is the gold standard investigation** to locate the lesion and to take biopsy which is very much required for further management of the case (Key Box 40.2).

Fig. 40.4: IVU showing filling defect

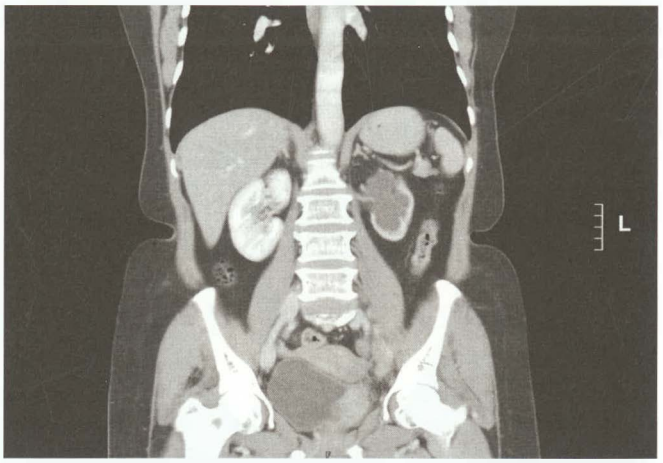


Fig. 40.5: CECT abdomen and pelvis showing a large tumour ir the left lateral wall of bladder with left hydroureteronephrosis

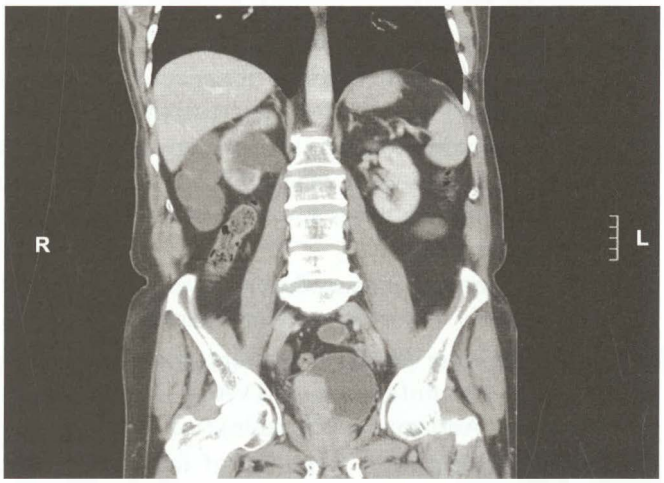


Fig. 40.6: CECT abdomen and pelvis showing a large tumour in the right lateral wall of bladder with right hydroureteronephrosis

Fig. 40.7: Ultrasound suggesting carcinoma urinary bladder

Fig. 40.8: Ultrasound suggesting carcinoma urinary bladder (another case)

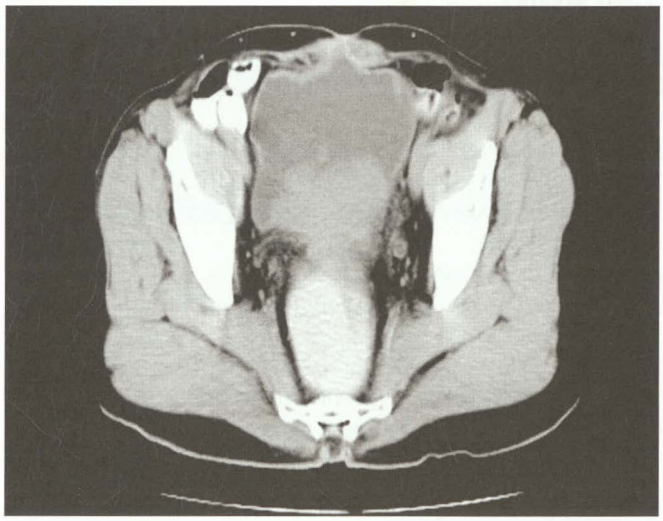


Fig. 40.9: Plain CT of the bladder showing filling defect in the bladder

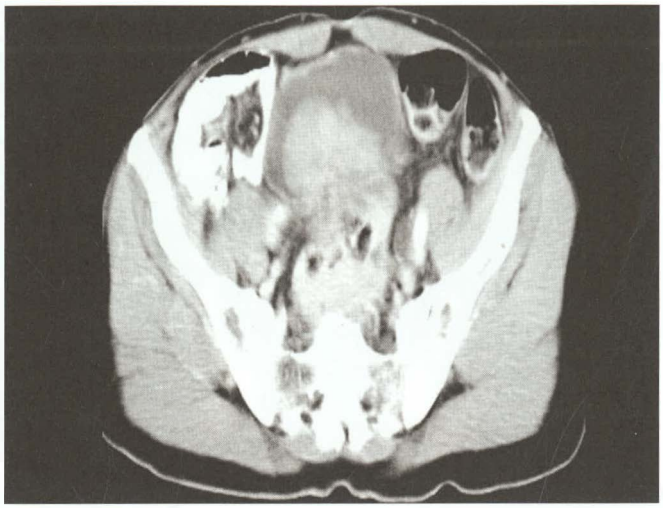


Fig. 40.10: Contrast CT of the bladder showing filling defect suggestive of carcinoma bladder

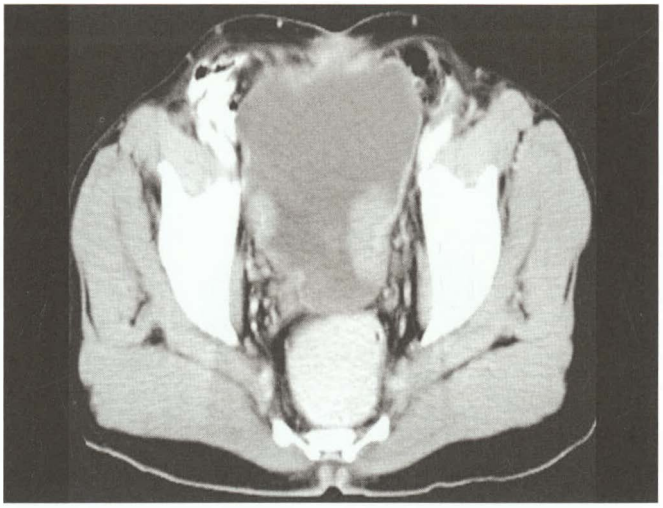


Fig. 40.11: Contrast CT of the bladder showing filling defects in two places suggestive of carcinoma bladder

KEY BOX 40.2

INDICATIONS FOR CYSTOSCOPY

- Haematuria with normal IVP
- Lower urinary tract symptom
- Malignant cells in the urine cytology.

6. Bimanual palpation, rectoabdominally in males and vaginoabdominally in females is done under general anaesthesia. Thickening of bladder wall, mobility, fixity and hardness can be made out.

Staging of bladder cancer

1. Clinical staging: Jewett, Strong and Marshall system

- Clinically the tumours are broadly classified into three groups: Superficial or noninvasive, infiltrating or invasive and carcinoma *in situ*.

2. TNM staging

TNM STAGING TNM staging of carcinoma bladder

Tis	–	Tumour *in situ*
Ta	–	Tumour involving mucosa without invading the lamina propria
T1	–	Tumour involving mucosa, invading lamina propria and submucosa
T2	–	Tumour involving the muscle layer
T3a	–	Tumour involving the muscle layer throughout the thickness
T3b	–	Tumour extending to the perivesical fat or peritoneum and involving the adjacent organs
T4	–	Involvement of the rectum and prostate
N0	–	No lymph nodes
N1	–	Lymph nodal metastasis
M0	–	No distant metastasis
M1	–	Distant metastasis present

Treatment of carcinoma urinary bladder

I. Carcinoma not involving muscle layer (Tis Ta, T1)

a. Transurethral resection (TUR) of tumour (resected base to be screened for tumour by microscopy).

b. Postoperative intravesical chemotherapy with Thiotepa/Adriamycin/Mitomycin retained inside the bladder for 1 hour. Such 6–8 courses at weekly interval are given to reduce recurrence.

c. BCG or interferon immunotherapy given postoperatively intravesically to prevent tumour recurrence.

II. T2–T4 lesions: Radical cystectomy followed by systemic chemotherapy (MVAC: Methotrexate, Vinblastine, Adriamycin, Cisplatin) (*vide infra*).

Radical cystectomy: Removal of the bladder with pericystic fat and the prostate and seminal vesicles, urethra in men and bladder and pericystic fat, cervix, uterus, anterior vaginal vault, urethra and ovaries in women. It is a major surgery with 3 to 8% mortality rate.

III. Any T, N1, M0 or any T, N0, M1–**Systemic chemotherapy** (**MVAC**) followed by radiation therapy has to be given.

IV. Small lesion involving muscle in the vault of bladder or posterolateral wall of the bladder, **partial cystectomy** (segmental resection) of that part of the bladder containing the growth with a wide margin of 2–3 cm. This should be followed by intravesical chemotherapy.

Role of radiotherapy

I. Local: If lesion is not anaplastic, is 4 cm or less, after open diathermy excision, radiotherapy can be given.

a. Implantation of radioactive Gold grains—^{198}Au

b. Radioactive Tantalum wire—^{192}Ta

II. Deep X-ray therapy

- Indication: Undifferentiated carcinoma
- By using Cobalt 60 or linear accelerator.

ECTOPIA VESICAE (EXOSTROPHY OF THE BLADDER)

- A rare congenital anomaly seen in 1: 50,000 births
- Male: Female = 4 : 1

Aetiopathogenesis

- This occurs due to failure of development of lower abdominal wall and anterior wall of the urinary bladder.
- As a result, the posterior bladder wall is seen protruding out below the umbilicus. **Hence, it is exostrophy of the bladder.**

Types

1. **Complete:** Pubic symphysis is not formed, complete epispadias in male or bifid clitoris in female.
2. **Incomplete:** Pubic symphysis, penis or clitoris are normal.

Clinical features (Key Box 40.3)

- More common in male children
- **Posterior bladder wall is seen in the lower abdomen** as a pink to red mucosa, partially inflamed.
- Umbilicus is usually absent.
- **Penis is rudimentary** and epispadias may be present.
- Testis descends normally into a well developed scrotum.
- **Pubic symphysis is widely separated.** It has the advantage in female patients in that it facilitates the delivery.
- In **female children—umbilicus is absent**, external genitalia are poorly developed, and clitoris is bifid.
- Constant dribbling of urine outside—therefore, they smell of urine.
- Recurrent urinary tract infection (UTI).

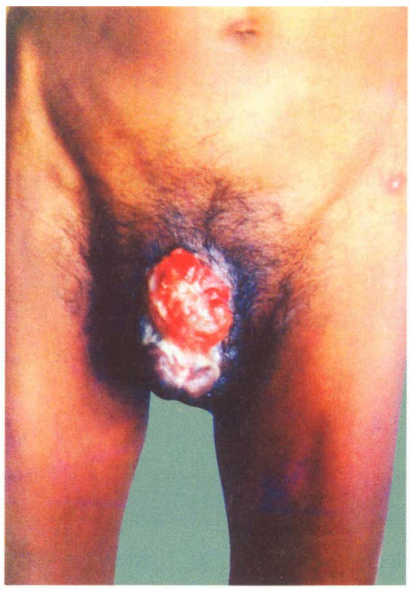


Fig. 40.12: Carcinoma bladder in a case of ectopia vesicae. (*Courtesy:* Dr PS Aralikatti , Associate Professor, BIMS, Belgaum, Karnataka)

Complications

1. Renal failure due to recurrent UTI
2. Adenocarcinoma of bladder and in early age (Fig. 40.12)
3. Ammoniacal dermatitis of the skin.

KEY BOX 40.3

ECTOPIA VESICAE

- Penis is rudimentary
- Posterior bladder wall is seen in lower abdomen
- Pubic bone is widely separated
- Poorly developed external genitalia
- Poor health—UTI, renal failure, Ca bladder

Treatment

1. **Incomplete:** Reconstruction of the anterior wall of the bladder with reconstruction of the bladder sphincter.
2. **Complete:** Total cystectomy followed by urinary diversion by implantation of ureters in the sigmoid colon **(ureterosigmoidostomy)** followed by reconstruction of anterior abdominal wall if the patient has urinary incontinence.

ACUTE CYSTITIS

Aetiology and pathogenesis

Acute uncomplicated bacterial cystitis predominantly affects women. By definition these are infections occurring in the **absence of any anatomic or functional abnormality** of the urinary tract. The ascending faecal—perineal—urethral route is the primary source of infection. Men are somewhat protected from ascending infection because of long urethra and antibacterial properties of prostatic secretions.

Causative organisms

80% of bladder infections in women are caused by *E. coli* followed by other gram-negative organisms such as *Klebsiella* and *Proteus* species.

Clinical features

Irritative voiding symptoms (frequency, urgency, dysuria) are the hallmarks of cystitis. Low backache and suprapubic pain are other complaints. Fever and other constitutional symptoms are usually present. Physical examination is frequently unremarkable except for suprapubic tenderness.

Diagnosis

- **Urinary microscopy** is the mainstay of diagnosis. Diagnosis is strongly considered positive if microscopy shows **> 5 WBCs**/high power field in females and 2–3 WBCs/high power field in males.
- **Urine culture** not only confirms the diagnosis but also identifies the causative organisms.

- Other tests and imaging studies are not indicated in an uncomplicated infection, unless patient presents with recurrent episodes.

Management

- Antibiotic therapy based on the culture and sensitivity report given for a period of 7–10 days, is curative.
- Symptomatic treatment in the form of antipyretics, urinary analgesics and antispasmodics may help.

DIVERTICULA OF THE BLADDER

Types

- **Congenital (situated midline anterosuperiorly):** Rare and usually symptomless. They represent the **unobliterated vesical end of the urachus**. May require excision if chronic infection persists.
- **Acquired:** They are pulsion diverticula and occur due to **bladder outflow obstruction.**

Pathology

- The diverticulum is lined by bladder mucosa
- The opening (mouth) is situated above and to the outer side of one ureteric orifice.

Clinical features—most commonly in males (95%) >50 years of age

- **Symptoms of recurrent urinary infection:** Suprapubic pain, frequency of micturition, fever with chills, etc.
- **Symptoms of lower urinary obstruction:** Frequency, urgency, hesitancy, etc.
- **Symptoms of pyelonephritis:** Backache, fever and renal angle tenderness, etc.

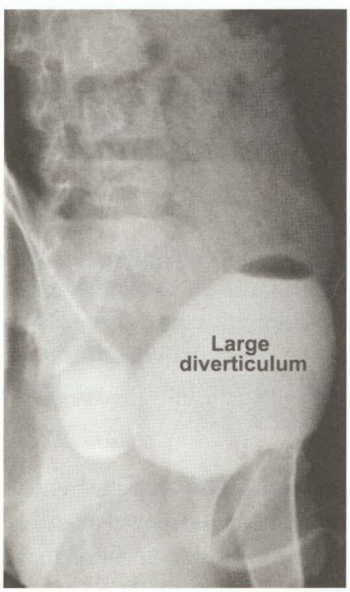


Fig. 40.13: Bladder diverticulum arising from the posterior wall. Ureter is seen entering the diverticulum

PEARLS OF WISDOM

Presence of diverticula is not an indication for surgery

Investigations

1. **Cystoscopy:** Full bladder distension is necessary to search for diverticulum.
2. **Intravenous urography:** It can detect the site of diverticulum. It can also detect hydronephrosis (Fig. 40.13).
3. **Ultrasonogram**
   - It can detect residual urine
   - It can detect diverticulum
   - It can detect associated stone

Treatment

Combined intravesical and extravesical diverticulectomy

Complications (Key Box 40.4)

KEY BOX 40.4

COMPLICATIONS

- Recurrent urinary infections
- Bladder stone can occur and it may give rise to haematuria
- Hydronephrosis and hydroureter occur due to peridiverticular inflammation and fibrosis
- Neoplasm: Squamous metaplasia and leukoplakia

URINARY FISTULAE

Introduction

Urinary fistulae are not an uncommon problem encountered by surgeons. Broadly classified into **congenital** and **acquired.** Congenital causes are a few of them (Key Box 40.5) which have been discussed in more detail in their respective chapters. More important ones are acquired fistulae which are given below. Among these vesicovaginal fistula is discussed in more detail.

Acquired fistulae

1. Traumatic urinary fistula

Perforating wounds, penetrating wounds or following surgery in the pelvis.

KEY BOX 40.5

CAUSES OF CONGENITAL URINARY FISTULAE

- Ectopia vesicae
- Patent urachus
- Association with imperforate anus

2. Vesicovaginal fistula

Causes

- Protracted or neglected labour
- Gynaecological operations such as total hysterectomy and anterior colporrhaphy
- Radiation causing avascular necrosis of the bladder
- Carcinoma cervix infiltrating into the bladder.

PEARLS OF WISDOM

Leakage due to necrosis of tissues manifests usually after 7 days.

Clinical features

- Leakage of urine from vagina
- Excoriation of the vulva

Diagnosis

- Digital vaginal examination may reveal thickening on the anterior wall of vagina.
- **Vaginal speculum examination:** Dribbling of urine into vagina.
- **Swab test:** Methylene blue is injected into the urethra and if vaginal swab is coloured blue, it is vesicovaginal fistula.

Treatment

- **Low fistula:** Transvaginal repair
- **High fistula:** Suprapubic approach and repair.

3. Fistula from renal pelvis to skin or gut

- Tuberculosis causes caseation and may result in fistula in the loin
- Large staghorn calculi
- Pyonephrosis
- Crohn's disease of the renal pelvis.

INTERSTITIAL CYSTITIS

- Initial symptoms are increased frequency and pain is relieved by micturition and aggravated by overdistension of bladder.
- First described by Guy Hunner (Gynaecologist) in 1914.
- The characteristic linear bleeding ulcer is caused by splitting of mucosa when the bladder is distended under anaesthesia.
- It is also called **Hunner's ulcer**.
- It is common in western female patients. Many of them are psychiatric patients.
- There is severe fibrosis of the urinary bladder due to pancystitis, resulting in a **small thimble bladder**. (In India, tuberculosis must be considered.)
- Frequency of micturition and pain due to decreased bladder capacity are the features. It causes sterile pyuria.
- Cystoscopy and biopsy will confirm the diagnosis
- Treatment is difficult—hydrostatic dilatation, instillation of dimethyl sulphoxide or surgical procedures such as ileocystoplasty have been tried.

SCHISTOSOMA HAEMATOBIUM

- Commonest cause of calcification in the bladder wall.
- It is called as urinary bilharziasis.
- The disease is caused by embryos (cercariae) of schistosoma, which enter the body through penetration of the skin and reach the bladder *via* the portal vein in a retrograde manner. In the bladder, ova are released which are excreted back into the fresh water *via* the urine. Fresh water snail is the intermediate host.
- Multiple pseudotubercles, nodules, granulomas, fibrosis are the **prominent pathological** features.
- Diagnosis is suspected by painless terminal haematuria, which lasts for 5 days (swimmer's itch) fever and eosinophilia.
- Cystoscopy and biopsy to confirm the diagnosis.
- Treated by long-term praziquantel and surgery may be required (ileocystoplasty).

PEARLS OF WISDOM

Urinary bilharziasis is a premalignant condition.

URINARY DIVERSION

Patients with lower urinary tract cancers or severe functional or anatomic abnormalities of the urinary bladder may require urinary diversion.

The most commonly used method of urinary diversion is by incorporating various intestinal segments into the urinary tract. Virtually every segment of the intestinal tract has been used.

1. **Ileal conduit:** 18–20 cm of ileum is used as a conduit. Ureters are directly implanted into it. The end of the ileal conduit is brought through lateral aspect of rectus abdominis muscle and stoma is made.
   This simply acts as a conduit carrying urine from the renal pelvis or ureter to the skin, where urine is collected in an appliance attached to the skin surface. It is not a continent mechanism.
2. **Ureterosigmoidostomy** is an example of continent urinary reservoir, wherein ureters are anastomosed into the sigmoid colon.

PEARLS OF WISDOM

The most worrisome complication of this procedure is development of adenocarcinoma, at the site where ureters are implanted.

- Routine sigmoidoscopy is recommended annually, to be started after 5 years of procedure.
- Newer method of continent diversion—orthotopic bladder substitution.

3. **Nephrostomy:** It is required for drainage and decompression of the upper urinary tract and is indicated in following situations:
   - Retrograde ureteral catheterisation is not advisable (e.g. in sepsis secondary to ureteral obstruction).
   - Retrograde ureteral catheterisation is impossible (e.g. complete ureteral obstruction by stone, tumour or stricture).
   - It is done by percutaneous approach.
4. **Uretero-ureterostomy:** It means anastomosis of ureter to ureter following resection. It can be on the same side, if the resection length of the ureter is 4 cm or less or to the opposite side ureter, if the ureter excised is more than 4 cm in length.

Indications

a. Trauma to ureter

b. Ureteric involvement by neoplastic conditions, e.g. colonic carcinoma, which requires resection of ureter.
   - Mainly indicated for upper and midureteral involvement. The procedure of choice for lower ureteric involvement is reimplantation into the bladder.

RUPTURE OF THE URINARY BLADDER

Causes of urinary bladder rupture

1. **Surgical** (iatrogenic): Bladder can be injured mostly during pelvic surgery, i.e. excision of the rectum or during gynaecological procedures, etc.
2. **Trauma:** Blunt injury abdomen due to road traffic accidents.
   - Kick or blow on abdomen, with a full bladder
   - Penetrating injury (very, very rare).

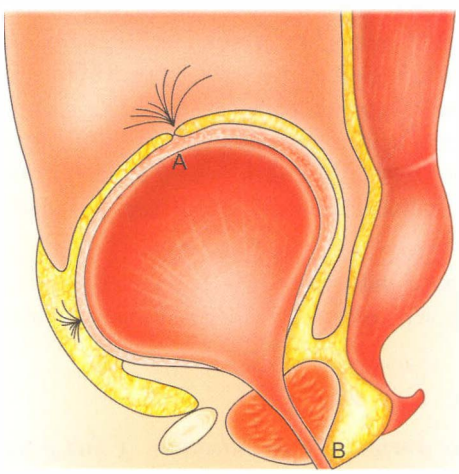


Fig. 40.14: Rupture bladder: (A) Intraperitoneal; (B) Extraperitoneal

Types of rupture and clinical features

I. Intraperitoneal rupture

When there is surgical trauma or trauma to a distended bladder the rupture will be intraperitoneal (Fig. 40.14A).

Clinical features

- Sudden severe suprapubic pain, hypotension/syncope and shock.
- Lower abdominal guarding and rigidity occur after a few hours of injury.
- Distension
- Even though the patient has not passed urine for a few hours there is no desire to micturate.
- Shifting dullness may be elicitable.

II. Extraperitoneal rupture (Fig. 40.14B)

- Trauma either penetrating or blunt injury with fracture of pubis gives rise to this type of injury.
- Difficult to distinguish clinically from an injury to the membranous urethra.

Investigations

1. **Plain X-ray abdomen:** Lower abdomen shows ground glass appearance.
2. **IVP:** Extravasation of the dye into the peritoneal cavity or extraperitoneally.
3. **Retrograde cystourethrogram:** Confirms the site of leak.
4. However, CT cystogram is the investigation of choice today.

Treatment

1. **Intraperitoneal rupture:** Laparotomy, repair of the bladder in two layers with vicryl. Drain the suprapubic space with a tube drain. An indwelling urethral catheter has to be placed for 10 days to 2 weeks to keep the bladder decompressed.
2. **Treatment of extraperitoneal rupture:** Extraperitoneally expose the bladder with a suprapubic midline incision and repair of the bladder. Drainage, as mentioned above, has to be carried out.

SURGICAL ANATOMY OF THE URETHRA (Fig. 40.15)

- Male urethra is divided into anterior urethra (*bulbopenile*) and posterior urethra (*prostatomembranous* urethra).
- Male urethra **functions as a conduit for urine and semen**. Anterior urethra is covered with erectile tissue of corpus spongiosum. Anterior urethra penetrates the urogenital diaphragm to enter the pelvic cavity as prostatomembranous urethra.
- Since its margins are attached to the perineal membrane it is **vulnerable to tear** at this **point in pelvic bone fracture**.
- Length of male urethra is about 14–16 cm.
- Entire urethra is supplied by internal pudendal artery.

- Veins drain into **Santorini's plexus** around the bladder neck and prostate.
- Female urethra is short, drains only urine and is not vulnerable for injuries.
- Narrowest part of male urethra is the external meatus.
- Prostatic urethra has 2 sphincters at each end. Internal sphincter at the bladder neck is composed of smooth muscle fibres. **The external sphincter is a rhabdosphincter**, about 2 cm long, surrounding the membranous urethra.
- Normally, urinary continence is maintained by the external sphincter. When the external sphincter is damaged (e.g. trauma, postsurgical) the internal sphincter maintains continence but to a lesser degree.
- Continence is not affected by ablation of internal sphincter (e.g. post-transurethral resection of prostate (TURP) but it results in **retrograde ejaculation**, i.e. the semen goes back into the bladder instead of exiting through the urethra.

RUPTURE URETHRA

Types

I. Rupture bulbar urethra

- The most common urethral injury
- Urethra angulates in the perineum, where it gets injured.
- Superficial extravasation of urine.

Clinical triad of rupture of bulbar urethra

1. Perineal haematoma
2. Urethral haemorrhage: Blood at the urethral meatus
3. **Distended bladder:** Diagnosed by percussion over suprapubic region which gives dull note.

Treatment

- **Advise the patient not to try to pass urine.**
- Urinary antibiotics
- Should be shifted to the operation theatre and with aseptic precautions a **catheter is passed gently**, if it enters the bladder, it is kept in place **for 2 weeks** and **perineal haematoma is drained.**
- If catheter does not reach the bladder, an incision is made in the perineum and the catheter is guided into the urethra.
- If all the measures fail, **emergency suprapubic cystostomy/catheter** is done to drain the urine and later repair of urethra is undertaken.

II. Rupture membranous urethra: Extraperitoneal rupture of the bladder

- Always (70%) associated with fracture of the pelvis.
- Occurs in major road traffic accidents.
- There may be disruption of pelvic bones, fracture symphysis pubis, with avulsion of the puboprostatic ligament leading to floating prostate.
- Deep extravasation of urine.

Types of rupture of membranous urethra

1. **Complete transection** results in floating bladder. In t condition, urethra is completely transected at the apex the prostate. As the puboprostatic ligament is avulsed, prostate falls back and migrates upwards. On rec examination prostate is felt as though it is floating **Floating prostate (Vermooten's sign).**
2. **Incomplete transection**
3. **Associated with injury to bladder:** Extraperitoneal ruptu of the bladder is seen here (intraperitoneal rupture of bladd occurs in a distended bladder).

Clinical features

- History of injury
- Features of shock due to significant blood loss (arour 1–2 litres)
- Haematuria
- In cases of extraperitoneal rupture of the bladder, it wi not be palpable due to extravasation of urine into perineum
- Suprapubic tenderness and dullness
- **Rectal examination:** Floating prostate can be felt whicl is tender.

Investigations

1. X-ray pelvic bones may show a fracture or separation o pubic symphysis.
2. Ascending urethrography (ASU) to confirm the rupture (Figs 40.16 and 40.17).
3. Once the stricture develops, voiding cystourethrogram (VCUG) is done to know the exact location of the stricture.

Treatment (Figs 40.18 and 40.19)

- Urgent blood transfusion to treat shock
- Suprapubic cystostomy is done and degree of damage assessed.
- A bougie/sound is passed from above and another similar sound is passed through the external meatus (penis). When the two meet, a click is appreciated. With both sounds in contact, the sound from bladder is withdrawn slowly. The lower one is advanced at this stage and 2nd sound appears in the bladder. A red rubber catheter is tied to it and the sound is withdrawn through external meatus.
- To this red rubber catheter which is seen outside, a Foley catheter is tied and is drawn into the bladder and is kept in place for 15 days. This is called **rail roading technique**.
- Associated injuries such as rupture bladder are treated by suturing.
- Antibiotics are given

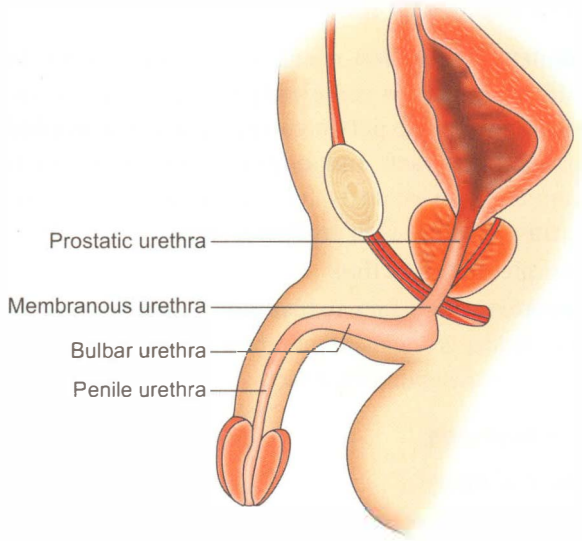


Fig. 40.15: Surgical anatomy of the urethra

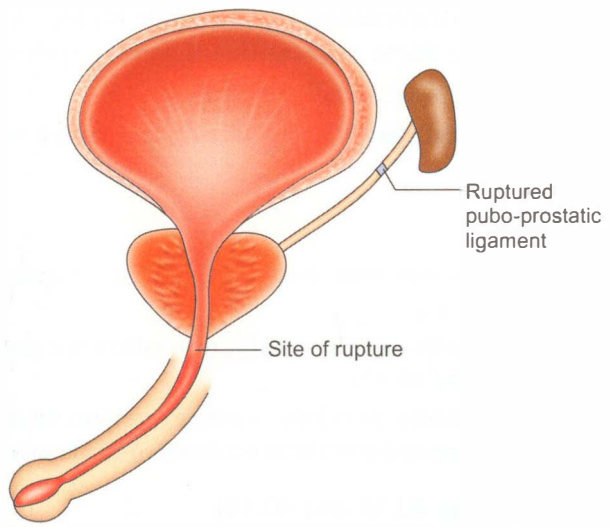


Fig. 40.16: Rupture of membranous urethra with floating prostate. Note the ruptured puboprostatic ligaments

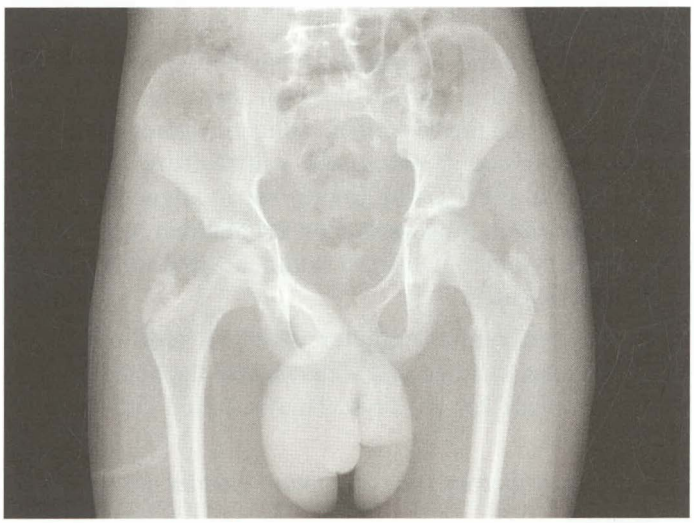


Fig 40.17: Plain X-ray showing extravasation of the dye

KEY BOX 40.6

COMPLICATIONS OF RUPTURE URETHRA

1. Extravasation of urine into scrotum, beneath superficial fascia of the penis, beneath Scarpa's fascia
2. Urethral stricture
3. Haematoma
4. Recurrent urinary tract infection

Complications of rupture urethra

Most dangerous complication is stricture urethra (Key Box 40.6)

STRICTURE URETHRA

Causes (Figs 40.20 to 40.22)

1. **Congenital**—very rare
2. **Post-inflammatory**
   - **A. Post-gonococcal urethritis**
     - Within 48 hours of exposure to the venereal disease gonorrhoea, there is involvement of periurethral glands. They are concentrated more in the bulbar urethra. Hence strictures are more in bulbar urethra.
     - It causes periurethral fibrosis, resulting in multiple dense strictures within 1 year of infection but may not cause difficulty in micturition for 10–15 years (Key Box 40.7).
   - **B. Tuberculosis**
3. **Post-instrumentation**
   - Catheterisation
   - Dilatation
   - Transurethral procedures
4. **Postoperative**
   - Prostatectomy
   - Repair of rupture urethra
5. **Rupture urethra**
6. **Schistosomiasis**

KEY BOX 40.7

ACUTE GONOCOCCAL URETHRITIS

- Pain during micturition
- Burning micturition
- **Gleet:** White flakes due to desquamated urethral epithelium in the early morning urine

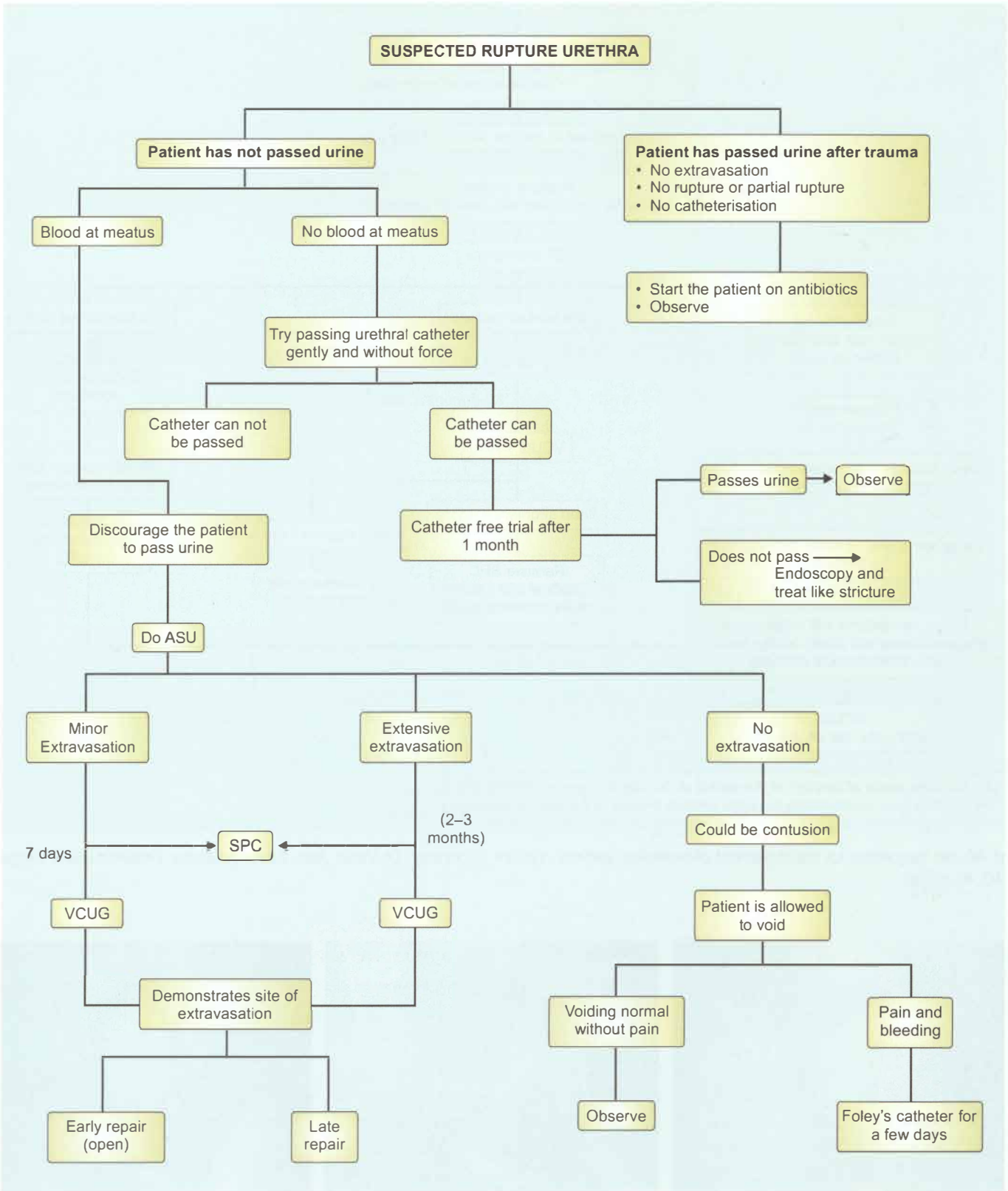


Fig. 40.18: Algorithm for management of bulbar urethral rupture (*Courtesy:* Dr Vikas Jain, Asst Professor, Department of Surgery, KMC, Manipal, 2007–2008)

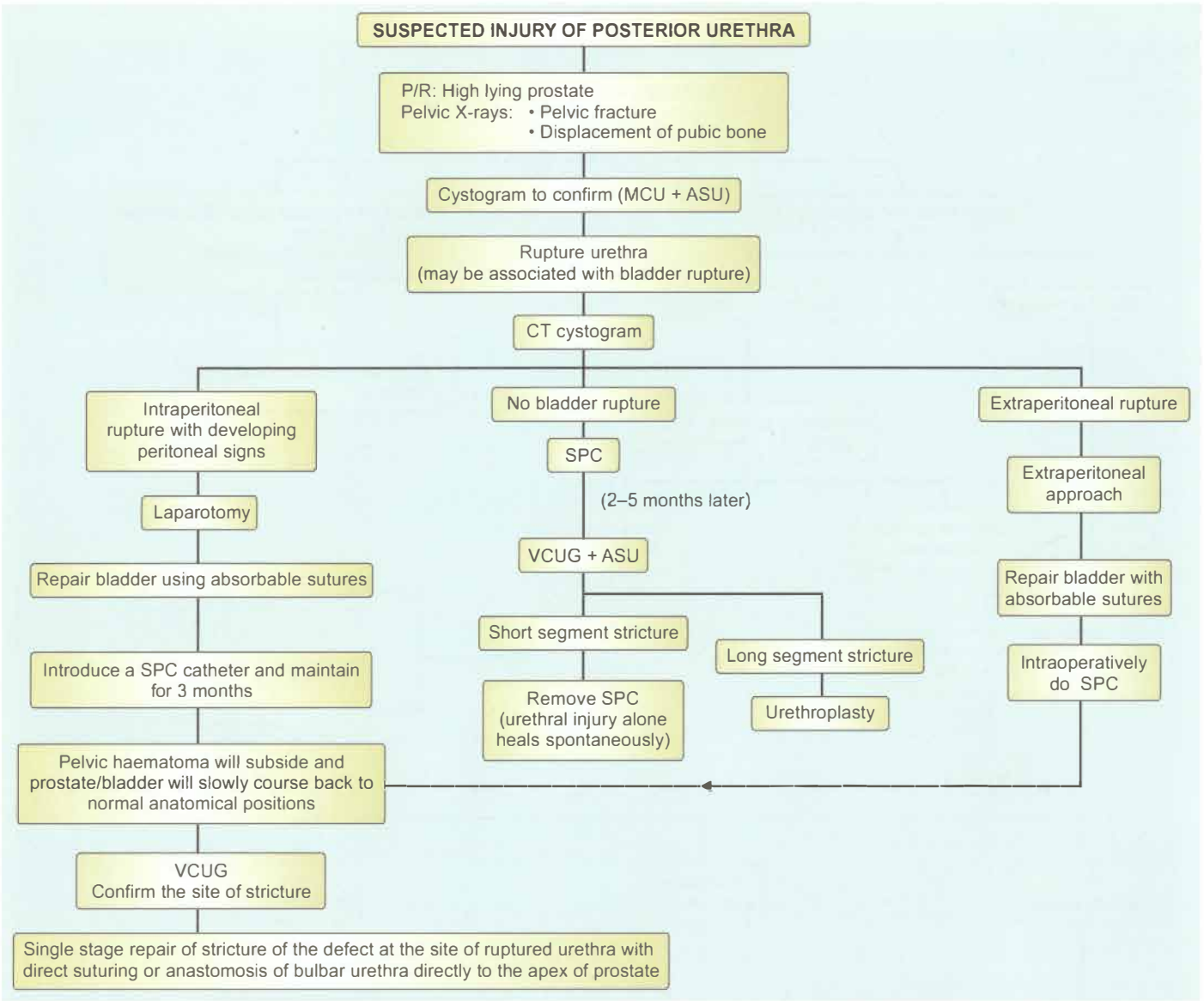


Fig. 40.19: Algorithm for management of posterior urethral rupture (*Courtesy:* Dr Vikas Jain, Asst Professor, Department of Surgery, KMC, Manipal)

Fig. 40.20: ASU and VCUG showing stricture urethra

Fig. 40.21: ASU: Total cut off seen due to stricture urethra

Fig. 40.22: ASU with MCU showing stricture

Clinical features

Previous history of exposure to gonorrhoea, history of instrumentation or history of injury to urethra is usually present.

- Common in young age (20–40 years).
- History of straining while passing urine.
- Suprapubic pain and swelling due to distended bladder.
- Stricture urethra may be felt in the perineum as a button hole.

It should be remembered that gonococcal urethritis is not common nowadays because of effective treatment of the disease.

Treatment

Usually cut at 12 o' clock position.

1. **Visual internal urethrotomy (VIU)** by using urethrotome
2. **Open method** is indicated in long strictures not responding to less invasive procedures. They are grouped under urethroplasty.
   a. Excision and end-to-end urethroplasty
   b. Reinforcement urethroplasty—buccal mucosa
   c. Substitution urethroplasty—buccal mucosa, skin
   d. Two step urethroplasty
3. **Regular dilatation** by using Lister's dilators

Complications

1. Acute retention of urine either following alcohol or due to postponement of micturition.
2. Secondary stones due to stasis of urine proximally.
3. Recurrent periurethral abscesses (multiple) which rupture and open externally in the perineal skin. When such a patient is asked to pass urine, urine can be seen coming out of multiple holes in the perineum **(Watercan perineum)**.
4. Recurrent epididymo-orchitis.

HYPOSPADIAS

In this condition, some portion of distal urethra is not developed, as a result of which external meatus is situated in the under surface of penis. Usually this is associated with chordee and hooded prepuce.

Types (Fig. 40.23)

1. Glandular variety

- In this external meatus is situated few mm away from normal site within the glans.

2. Coronal variety

- It occurs due to failure of development of urethra which runs in the glans penis.
- As a result of this, urethra opens at the corona glands-junction of glans and shaft of penis.

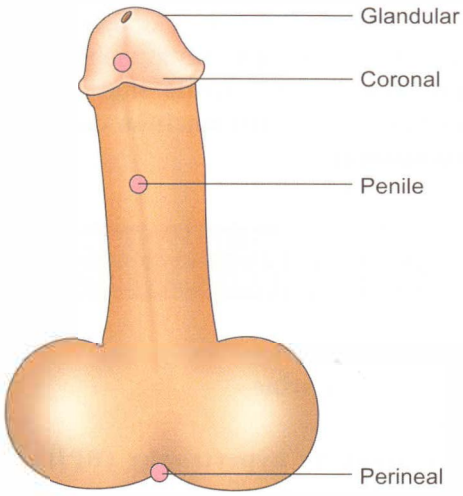


Fig. 40.23: Various types of hypospadias

- Both these varieties do not give major problems functionally. It can be left alone without treatment.

3. Penile hypospadias

- In this the external opening is situated somewhere in the under surface of the penis.

4. Peno-scrotal/perineal hypospadias

- In this condition, the entire urethra is not developed
- Penis is rudimentary.
- Urethral opening is seen in between two halves of the scrotum and often it is split.
- Cases may be associated with undescended testes
- In such cases it is difficult to differentiate the sex of the child

Clinical features

- Occurs in 1: 350 males.
- Micturition: Stream is good, but it wets the clothes in 3rd and 4th varieties.
- Chordee: Many of the cases are associated with bending of penis.
- Sexual intercourse will be difficult.
- Hooded prepuce.

In severe hypospadias, possibility of intersex problem is settled by **karyotyping.**

Treatment

1. One-stage urethroplasty

- Chordee correction: Always confirm by inducing artificial erection
- Urethral tube formation by tubularising urethra
- Inner prepuceal island tube urethroplasty.

2. Two-stage urethroplasty

- When the child is 6–12 months old, chordee is corrected by straightening the penis. This is called orthoplasty.

- When the child is 5–6 years old, reconstruction of the urethra is done by using locally available skin either from the prepuce or from penile shaft. This is called urethroplasty. Hence, **circumcision should not be done in hypospadias.**

DIFFERENTIAL DIAGNOSIS OF RETENTION OF URINE

Causes of retention of urine

I. Acute retention of urine

A. In males

- **Benign prostatic hypertrophy (BPH):** In elderly patients over 50 years of age.
- **Stricture** urethra: In young patients
- **Postoperative retention** of urine: Operations like haemorrhoidectomy, fistulectomy, etc. produces reflex spasm of internal sphincter, which precipitates retention of urine. Management of such cases is given in Key Box 40.8.

B. In females

- Hysteria
- Retroverted gravid uterus
- Urethral stenosis

Residual urine

Normal—30 ml
>50—abnormal/significant
>200 in BPH requires surgery

C. In children

- Meatal stenosis due to meatal ulcer with scab (due to scratching habit of child).

D. In general

- Spinal anaesthesia
- Spinal injuries
- Blood clot in the bladder following prostatectomy
- Bladder stone in school-going children: Pain referred to tip of penis
- Acute urethritis and acute prostatitis due to bacterial infection
- Faecal impaction in the rectum

KEY BOX 40.8

POSTOPERATIVE RETENTION OF URINE: TREATMENT

1. Hot water fomentation to the suprapubic region
2. Provide privacy
3. Run a tap nearby
4. Make the patient to stand and pass urine
5. Catheterisation should be done as the last resort

- Contracture of the bladder neck
- Urethral calculus
- Drugs: Atropine, carbachol, bethanechol.

II. Chronic retention of urine (Key Box 40.9)

1. Benign prostatic hypertrophy
2. Bladder neck contracture
3. Stricture urethra

KEY BOX 40.9

CHRONIC RETENTION OF URINE

- BPH: Most common cause
- Painless
- Suprapubic dullness
- Slow decompression is recommended

POSTERIOR URETHRAL VALVE

- They are **congenital, symmetrical valves** in the posterior urethra.
- It is one of the **common causes of vesicoureteric reflux** and hydronephrosis in infants.
- Bladder wall is thickened due to obstruction and hypertrophy. The urinary bladder is palpable, hard and felt in the suprapubic region—**cricket ball bladder**.
- Due to stasis, recurrent urinary tract infection occurs commonly.
- It is also one of the **common causes of renal failure** in infancy and childhood.

Investigations

- **Ultrasound:** Bilateral hydronephrosis, thickened bladder, etc.
- **Micturating cystourethrography:** Dilated proximal urethra is highly suggestive of posterior urethral valve. **It is a diagnostic investigation** (Figs 40.24 to 40.27).

Treatment

- Cystoscopic posterior urethral valve fulguration
- In very ill patients, initially vesicostomy is done to improve renal function and stabilise the patient. Fulguration is done after 1–2 weeks.
- These patients should be monitored for renal failure by serial creatinine measurements.

PEARLS OF WISDOM

It can be associated with anorectal and vertebral malformations.

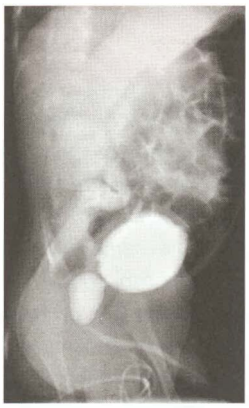


Fig. 40.24: PUV with grade IV reflux

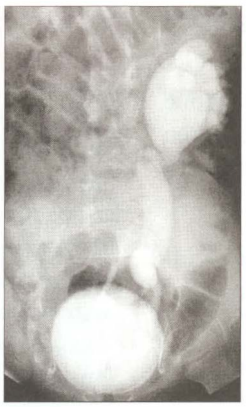


Fig. 40.25: MUC showing grade V vesicoureteric reflux

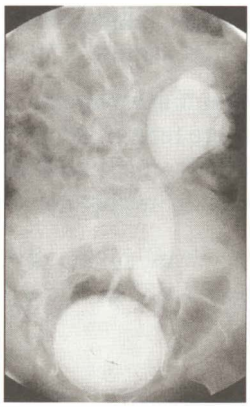


Fig. 40.26: Bilateral vesicoureteric reflux

Fig. 40.27: Micturating cystourethrogram showing bilateral grade V vesicoureteric reflux (VUR)

VESICOURETERIC REFLUX (VUR)

Vesicoureteric reflux is the retrograde flow of urine from the bladder into the kidneys.

Normally urine flows from the kidneys into the bladder and backward flow is prevented by a complex anatomy at the vesicoureteric junction, most importantly a good length of submucosal tunnel of ureter. Lack of adequate submucosal tunnel leads to VUR.

The VUR can be of mild to severe grades (graded I-mild to V-severe).

Clinical features

- Child might be diagnosed to have hydronephrosis detected antenatally, depending on the severity of reflux.
- **Recurrent UTI:** Due to increased amount of residual urine. When the child voids, some urine refluxes back into the kidneys, which comes back into bladder after voiding ceases. This will lead to a high post-void residue (PVR), causing recurrent infections.
- **Pyelonephritis and scarring:** Reflux of infected urine causes pyelonephritis. It can be acute or chronic. Recurrent episodes can lead to scarring of kidneys and decreased function, proteinuria and hypertension. Severe scarring bilaterally can lead to chronic renal failure (CRF) and end-stage renal disease (ESRD).

Investigations

- Urine analysis for proteinuria and infection.
- Complete blood picture: Anaemia in CRF and leukocytosis in acute pyelonephritis.
- Renal function tests: Increased urea and creatinine in renal failure.
- Ultrasonography (USG): Renal size, parenchyma and hydroureteronephrosis with PVR can be assessed.
- **Micturating cystourethrogram (MCU)** or voiding cystourethrogram (VCUG)—**investigation of choice** for diagnosis. Procedure: Bladder is filled with contrast after catheterisation and X-rays taken during filling and full bladder to look for reflux. After full bladder, catheter is removed, patient is asked to void and X-rays are taken in the voiding phase.
- **DMSA scan**—for extent of renal scarring and function. Helps in choosing the management strategy (medical vs surgical).

Treatment

I. Conservative management: Low grade reflux (grades I–III) can be managed conservatively. The aims of conservative therapy include:

- Prophylactic/suppressive antibiotics.
- Regular or timed voiding and double voiding to keep PVR as low as possible.
- To avoid constipation—reduces the incidence of UTI.
- With these measures, most low grade reflux resolves spontaneously.
- Higher grade reflux requires endoscopic or surgical management.

II. **Endoscopic management:** Injection of a bulking agent to provide support to VUJ, helps in a few cases.

III. **Surgical management:** Ureteric reimplantation is done for severe reflux. The principle of surgery is to lengthen the submucosal tunnel. If one kidney is poorly functioning, nephrectomy can be done.

In some cases, bilaterally scarred, nonfunctioning kidneys can lead to ESRD. These patients require renal transplantation.

MISCELLANEOUS

INVESTIGATION OF CHOICE

- In carcinoma bladder—cystoscopy
- In rupture bladder—CT cystogram
- In rupture urethra—ascending urethrography
- In posterior urethral valve—micturating cystourethrography (MCU).
- In vesicoureteric reflux—MCU or VCUG

CLINICAL NOTES (*see* Fig.40.3, page 955)

A 45-year-old multiparous female patient was diagnosed to have bladder stone. She underwent vaginal examination because she also had pain in her genitalia.She underwent dilatation and curettage.On dilatation, tail of Cu-T with thread was seen in the uterus.On trying to remove Cu-T, only thread could be extracted. She underwent laparotomy. Uterus was found plastered to urinary bladder. On opening the bladder, a stone of 6 × 6 cm with Cu-T in pregnated in it was present. The Cu-T had perforated anterior wall of uterus and posterior wall of the bladder. It was removed. (Courtesy: Prof Rajiv Shetty, Professor Shivaswamy, Professor Durganna, Bangalore Medical College, Bengaluru)

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All the topics have been updated
- Vesicoureteric reflux has been added
- New images are added

MULTIPLE CHOICE QUESTIONS

1. Following type of malignant tumour can occur in urinary bladder *except*:
A. Transitional carcinoma
B. Adenocarcinoma
C. Squamous cell carcinoma
D. Leiomyosarcoma

2. Following are true for internal sphincter of the urinary bladder *except*:
A. It is a smooth muscle
B. It is innervated by adrenergic fibres
C. It prevents retrograde ejaculation
D. It is supplied by pudendal nerve

3. Cystine calculi in the urinary bladder are radio-opaque because of:
A. High calcium content
B. High sulphur content
C. High triple phosphate content
D. High oxalate content

4. Following are clinical features of urinary bladder stone *except*:
A. Pain referred to the testis
B. Pain aggravated by jumping
C. Strangury
D. Haematuria

5. Which of the following parasitic infestations is a strong risk factor for carcinoma urinary bladder?
A. Bilharziasis B. Ascariasis
C. Leishmaniasis D. *Clonorchis sinensis*

6. Following are true for carcinoma urinary bladder *except*:
A. Aniline dye workers have high risk
B. Transitional cell carcinomas are more common
C. Strangury
D. Adenocarcinoma of the bladder is due to bilharziasis

7. Following are true in ectopia vesicae *except*:
A. Penis is normal
B. Posterior wall of the urinary bladder is seen
C. Pubic symphysis is widely separated
D. Adenocarcinoma urinary bladder occurs very early

8. Most common organism causing acute cystitis are:
A. *E.coli* B. Klebsiella
C. Proteus D. Pseudomonas

9. Following are the causes for vesicovaginal fistula *except*:
A. Protracted labour B. Anterior colporrhaphy
C. Radiation D. Crohn's disease

10. The most worrying complication following uretero-sigmoidostomy is:
A. Acidosis
B. Adenocarcinoma
C. Alkalosis
D. Recurrent infection and septicaemia

11. Which of the following is not the feature of intra-peritoneal rupture of the urinary bladder?
A. Shock
B. Urgent and frequent desire to pass urine
C. Suprapubic pain
D. Hypotension

12. Which is the narrowest portion of male urethra?
A. Penile urethra B. External meatus
C. Perineal urethra D. Bulbar urethra

13. Which of the following is not part of the triad of rupture of bulbar urethra?
A. Perineal haematoma
B. Blood at urethral meatus
C. Distended bladder
D. Floating prostate

14. What is the first advice given to the patient in rupture bulbar urethra?
A. Blood transfusion
B. Intravenous antibiotics
C. Not to try pass urine
D. Immediate catheterisation

15. Floating prostate—Vermooten's sign is classical of:
A. Rupture bulbar urethra
B. Rupture penile urethra
C. Rupture intraperitoneal urinary bladder
D. Rupture membranous urethra

16. Which of the following is true for posterior urethral valves?
A. They are acquired
B. Bladder is thin walled and more prone for rupture
C. Renal failure is uncommon
D. They are symmetrical valves

ANSWERS

1 D	2 D	3 B	4 A	5 A	6 D	7 A	8 A	9 D	10 B
11 B	12 B	13 D	14 C	15 D	16 D				

41

Prostate and Seminal Vesicles

- Surgical anatomy
- Structural anatomy
- Benign prostatic hyperplasia (BPH)
- Carcinoma of the prostate
- Gleason score
- Prostatitis
- What is new?/Recent advances

SURGICAL ANATOMY

Embryology and lobes

- The prostate develops around 12th week of intrauterine life. Primitive buds given from urethra form the glandular tissue and surrounding mesenchyme forms the fibromuscular stroma. Developmentally, the prostate has 5 lobes: Anterior, posterior, 2 lateral and 1 middle lobe.
- **Middle lobe (median lobe)** is situated in between the two ejaculatory ducts and the urethra. The enlargement of this lobe in benign hypertrophy of the prostate is responsible for the obstruction of urethra. This lobe enlarges upwards into the bladder (Fig. 41.1).

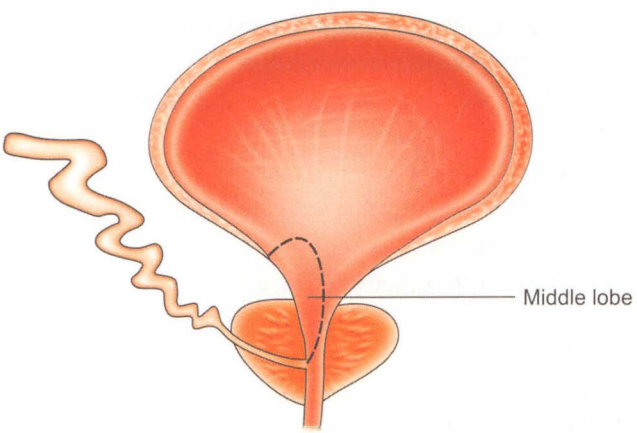


Fig. 41.1: Prostate—lobes

- In benign prostatic hypertrophy (BPH), the glands of the inner adenomatous zone hypertrophy and lead to urinary outflow obstruction. **Carcinoma usually occurs in the outer nonadenomatous zone.**
- New terminology for BPH arising zone is **transitional zone** and carcinoma arising zone is **peripheral zone**.

Structural anatomy

- Prostatic urethra is surrounded by a fibroadenomatous gland.
- Urethral glands open into the prostatic urethra. These **submucosal glands are responsible for BPH**.
- When the prostate enlarges, it compresses the outer zone resulting in a false capsule.
- The **outermost zone** is the zone of prostatic glands proper which is responsible for **carcinoma of prostate** (Fig. 41.2).

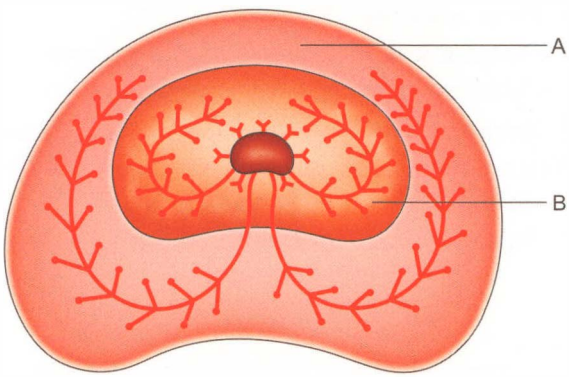


Fig. 41.2: Prostate: (A) Outer carcinomatous zone, (B) Inner adenomatous zone

- Surrounding this, there is **fascia of Denonvilliers** which is a part of pelvic peritoneum.
- Between the anatomical capsule and pelvic peritoneum, prostatic venous plexus is present which may give rise to massive haemorrhage, if injured.

BENIGN PROSTATIC HYPERPLASIA (BPH)

AETIOPATHOGENESIS

There are 2 theories to explain BPH.

1. Hormonal theory

- It has been compared to fibroadenosis in female patients.
- As the age advances, the levels of androgens come down. There is a corresponding increase in the oestrogen which stimulates the prostatic gland and produces BPH.

2. Neoplastic theory

According to this theory there is proliferation of all the elements of prostate: Fibrous, muscular and glandular resulting in fibromyoadenoma.

SECONDARY EFFECTS OF BPH

1. Urethral changes

- Urethra gets **compressed, elongated and gets converted into a narrow, longitudinal slit**.
- The effect is more with median lobe enlargement which is due to enlargement of subcervical glands.
- Lateral lobes enlarge when there is involvement of submucous glands.

2. Changes in the bladder (Fig. 41.3)

- As a result of obstruction, the bladder musculature undergoes hypertrophy. Very prominent thick bundles of the muscle can be seen, which are called fasciculations or **trabeculations**.

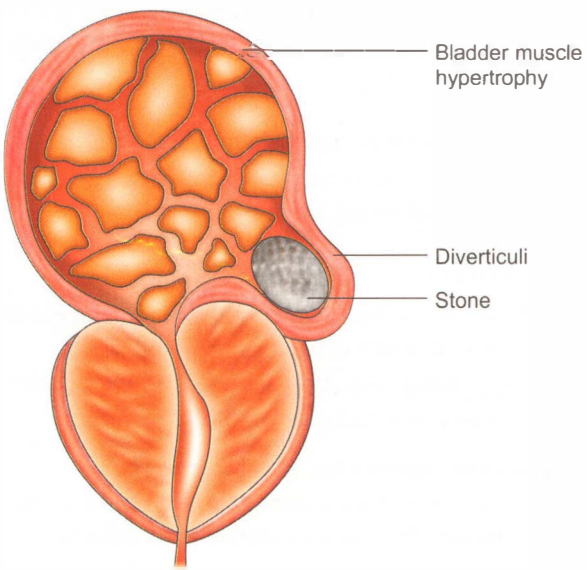


Fig. 41.3: Secondary changes in the urinary bladder due to BPH

- In between the fasciculations, there are depressed areas which are called **sacculations**.
- Since the sacculi are thin, as the pressure increases, herniation occurs outside resulting in diverticuli.
- In the diverticuli, there is stasis of urine resulting in secondary infection and stone formation.

3. Changes in the ureter and kidney (Key Box 41.1)

Bilateral hydronephrosis and bilateral hydroureter are the end result of BPH, which may result in renal failure.

KEY BOX 41.1

BPH—BACK PRESSURE EFFECTS ON THE BLADDER

Bladder muscle hypertrophy
↓
Fasciculations
↓
Sacculations
↓
Diverticuli
↓
Stasis, infection, stone

CLINICAL FEATURES OF BPH (Key Box 41.2)

- **Frequency, urgency, hesitancy are triad of BPH**
  - **Frequency:** To start with, frequency is present during the day time followed by day and night (5–10 times during the night). It is due to ineffective emptying of the bladder. It results in residual urine in the bladder precipitating cystitis.
  - **Urgency:** As the prostate enlarges there is **vesical introversion of sensitive mucous membrane of prostatic urethra within the bladder.** This causes the internal sphincter to stretch and prevents contraction. This results in a few drops of the urine trickling down the posterior urethra resulting in an urgent desire to pass urine (urgency).
  - **Hesitancy:** Patient hesitates to pass urine because it is so ineffective due to obstruction.

KEY BOX 41.2

BPH—CLINICAL FEATURES

- Frequency
- Urgency
- Hesitancy
- Acute retention of urine
- Chronic retention of urine

- **Haematuria is rare**
  - It is due to congestion of prostatic venous plexuses resulting in hyperaemia and haematuria.
- **BPH with acute retention of urine**
  - This occurs due to postponement of micturition, following alcohol or drugs like mydriatics.
- **BPH with chronic retention of urine**
  - Many of the patients present with **chronic retention** of urine, with painless enlargement of the urinary bladder.

COMPLICATIONS

- Stones
- Diverticuli
- Renal failure
- Recurrent urinary tract infection

DIAGNOSIS OF BPH

Digital rectal examination: Enlarged lateral lobes can be easily felt. Rectal mucosa is free (In an enlarged prostate gland, in case of carcinoma of prostate, the mucosa of the rectum cannot be moved if it has infiltrated into the rectum.).

Grading of prostate is done as follows

I. The prostatic lobes protrude minimally into the rectal lumen by 1–2 cm, the median sulcus is palpable.
II. Prostatic lobes protrude > 2 cm but < 3 cm into the rectal lumen and the median sulcus is obliterated.
III. 3–4 cm protrusion
IV. > 4 cm protrusion of lobes, most of the rectal lumen is filled by the projecting prostatic lobes.

INVESTIGATIONS

1. **Blood urea and creatinine:** Raised levels indicate renal failure.
2. **Uroflowmetry:** The person is asked to void urine from his full bladder into the flowmeter. The flow rate is assessed.
   Peak flow rate
   - Normal peak flow rate: 20 ml/sec.
   - Doubtful peak obstruction: 10 to 15 ml/sec.
   - Definite peak obstruction: Less than 10 ml/sec.
   - Thus, the degree of bladder outlet obstruction (BOO) can be secured by uroflowmetry in case of BPH.
3. **Ultrasonogram:** To assess the size and weight of prostate, to assess the residual urine and to look for hydro-ureteronephrosis, bladder wall changes.

TREATMENT

It can be classified into medical treatment and surgical treatment.

I. Medical treatment of BPH

If the patient has mere frequency of micturition and if the residual urine is not much (< 150 ml), uroflowmetry shows more than 15 ml/sec of urine flow and if there are no back pressure effects on the kidney, the patient can be reassured advised to avoid heavy alcohol consumption which may lea to prostatic congestion and acute retention of urine. To avoi over distension of the bladder, he has to void the urine as an when he feels the urinary sensation of micturition, and **shoul not postpone micturition**.

Drugs

a. **Finasteride acetate** 5 mg daily for 6 months. It is a 5α reductase inhibitor. **It helps in prevention of hyperplasi of the prostate. It is given for large prostates.**
b. **α-adrenergic blockers:** It is supposed to relax the interna sphincter for better drainage of the bladder. They are given when the prostate is less than 40 g (small) as measured by ultrasound. Tamsulosin (most selective), terazosin, alfazocin are a few drugs used today.

II. Surgical treatment of BPH

Indications for surgery

1. **Acute retention** of urine
2. **Chronic retention** of urine with postvoid residual urine more than 200 ml.
3. If **frequency** of micturition is so much that it disturbs the normal lifestyle during daytime.
4. **Complications** such as haematuria due to congestion of prostatic venous plexuses, hydroureteronephrosis, prostatic diverticulosis, vesical calculus and recurrent infections.

SURGICAL METHODS

1. Transurethral resection of the prostate (TURP)

- This is the most popular method today.
- A resectoscope is passed through the urethra and under vision with constant irrigation with water or glycine, the prostate is resected into multiple pieces and removed.
- Haemostasis is obtained with the help of a cautery.

Advantages

1. Postoperative recovery is smooth and rapid.
2. Incontinence is rare because the chances of damage to the internal sphincter are very low.

Disadvantages

1. TURP syndrome with water intoxication and electrolyte imbalance, if water is used as irrigating fluid.
2. If there is BPH with diverticuli and stone, TURP has to be followed by litholapaxy.

2. Transvesical suprapubic prostatectomy

- This method is now **restricted to glands more than 100 g in weight and associated with calculus**.
- Ankylosis of hip/other orthopaedic conditions (difficulty in positioning).

- Through an extraperitoneal approach the bladder is opened, prostate is enucleated with finger, bleeding is controlled by inflating the Foley bulb with about 30–50 ml of air and by ligatures.
- Bladder is drained by a Malecot's catheter which is wider than Foley's so that it can drain well if the bleeding occurs in the bladder.
- During the process, the prostatic urethra is also avulsed.
- After about 7–10 days, a tract develops along the length of Foley catheter which heals by granulation, fibrosis and forms the future prostatic urethra.

Disadvantages

- Blind resection
- Chances of haemorrhage are more
- Stricture of prostatic urethra

3. Retropubic prostatectomy

Done by extraperitoneal approach without opening the bladder, pushing the bladder to one side and excision of the prostate.

4. Perineal prostatectomy

Not done nowadays

Newer treatments

- Holmium: YAG laser.
- Intraurethral stents—in men who are grossly unfit (ASA Grade IV) for surgery.

CARCINOMA OF THE PROSTATE

- Carcinoma of the prostate is common after the age of 65 years. The incidence increases with age.
- In Western countries, it is **the second most common type of carcinoma in males after 65 years**, first being bronchogenic carcinoma.
- Prostatectomy done for BPH does not give protection against development of carcinoma of prostate because during prostatectomy the outer zone is left undisturbed (not resected).

CLINICAL FEATURES

1. Histological surprise

Prostatectomy done with diagnosis of BPH but histology reveals carcinoma of the prostate.

2. Multiple bone pain, confused for rheumatism, is due to multiple metastasis.

3. Rectal examination: Reveals a hard nodule on the anterior wall of the rectum, obliteration of median sulcus. The rectal mucosa cannot be moved over the prostate but it is not ulcerated (fascia of Denonvilliers prevents the spread of carcinoma prostate into the rectum).

4. Elderly man with **bilateral sciatica** with metastasis in the thoracolumbar vertebrae.

5. Acute retention of urine occurs in 5–10% of cases of carcinoma of prostate.

6. Difficulty in passing urine, painful micturition and sometimes with haematuria is due to involvement of prostatic urethra.

SPREAD (Key Box 41.3)

1. Haematogenous spread

- This is due to retrograde tumour embolisation which occurs through prostatic venous plexus which communicates through the emissary veins with the bone (Batson's paravertebral plexus of veins).

Peculiarities of secondary deposit from carcinoma of prostate:

- They are multiple
- Moth-eaten appearance
- Osteoblastic (in most of other secondaries, they are osteolytic).
- Most common site of origin for skeletal metastases.

KEY BOX 41.3

BONES INVOLVED IN CARCINOMA PROSTATE

1. Thoracolumbar vertebrae (Fig. 41.4)
2. Pelvic bone, iliac crest
3. Femur
4. Scalp
5. Ribs

2. Lymphatic spread

- Prostatic chain of lymphatics drain into internal iliac nodes.
- When spread occurs along seminal vesicle, external iliac nodes are enlarged.
- From this group of nodes, para-aortic nodes, mediastinal nodes, followed by left supraclavicular nodes get involved.

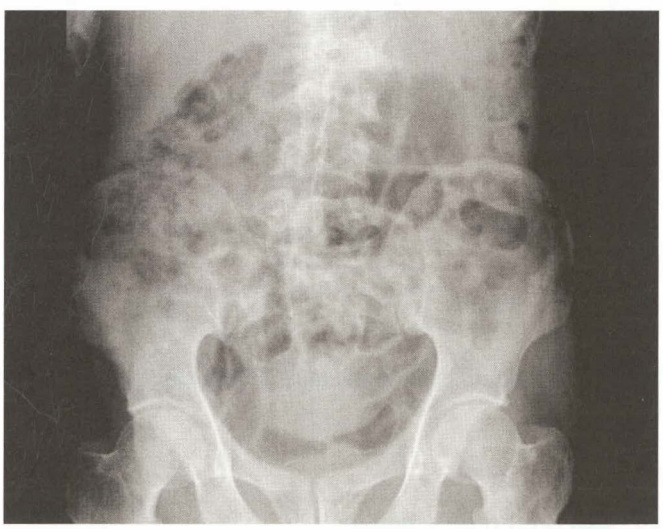


Fig. 41.4: Plain X-ray showing destruction of vertebrae

3. Local spread

- On the medial side it can involve the prostatic urethra and give rise to retention of urine.
- When it spreads upwards, the bladder can get involved resulting in painful haematuria.
- Superiorly it can also involve seminal vesicle.
- Rectum is involved very late in carcinoma of prostate because of the tough Denonvillier's fascia.

STAGING

TNM STAGING — Staging using the tumour, node, metastasis (TNM) system (Fig. 41.5)

1. **T1a, T1b and T1c.** These are incidentally found tumours in a clinically benign gland after histological examination of a prostatectomy specimen. T1a is a tumour involving less than 5 per cent of the resected specimen; these tumours are usually well or moderately well differentiated. T1b is a tumour involving > 5 per cent of the resected specimen. TIc tumours are impalpable tumours found following investigation of a raised PSA.
2. **T2a** disease presents as a suspicious nodule on rectal examination confined within the prostate capsule and involving one lobe. T2b disease involves both lobes.
3. **T3** tumour extends through the capsule (T3a, uni- or bilateral extension. T3b, seminal vesical extension).
4. **T4** is a tumour that is fixed or invading adjacent structures other than seminal vesicles—rectum or pelvic side wall

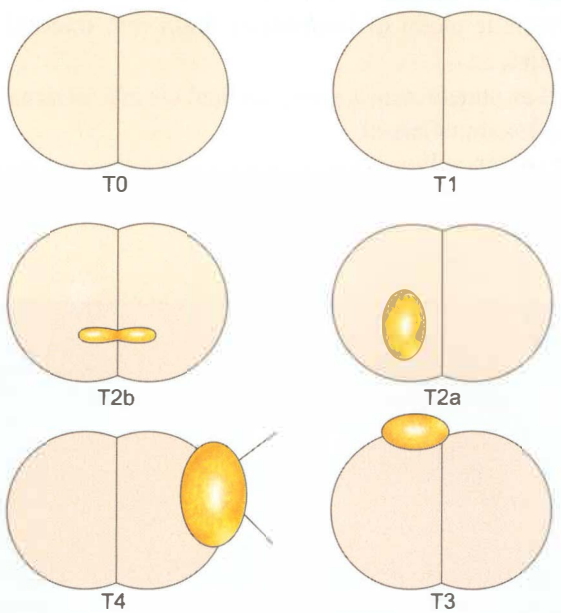


Fig. 41.5: Tumour, node, metastasis (TNM) staging for prostate cancer

INVESTIGATIONS

1. Transrectal trucut biopsy

Report—adenocarcinoma

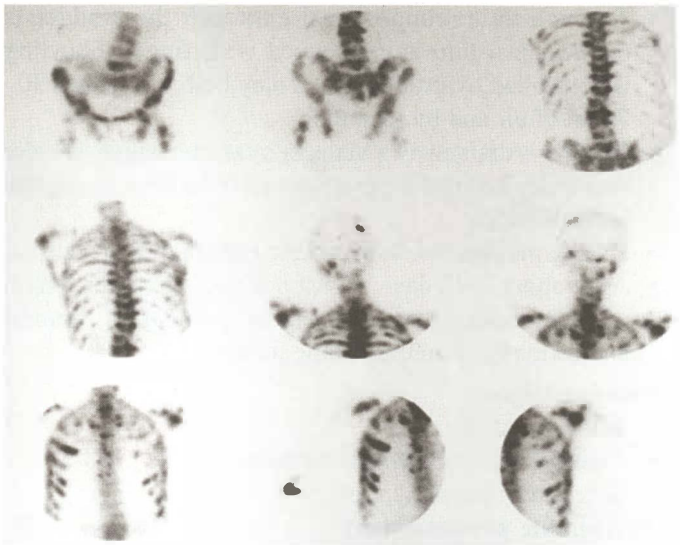


Fig. 41.6: Bone scan showing extensive metastasis

Fig. 41.7: Lytic lesions in pubic bones

2. X-ray of bones (Figs 41.6 and 41.7)

Which are likely to be involved (already mentioned).

3. Serum acid phosphatase (Key Box 41.4)

- The enzymes which split the organic phosphates are concentrated in the prostate which are responsible for acidic pH in the prostatic urethra.
- Normally they are drained in the urine so that they are not detectable in the serum.

KEY BOX 41.4

ACID PHOSPHATASE INCREASED IN

1. Paget's disease of bone
2. Acute prostatitis
3. Cirrhosis of liver
4. Carcinoma prostate

- In carcinoma prostate **because of the ductal blockage;** it gets absorbed into the blood and thus high levels are reached, especially with metastasis.
- 1 to 3 King-Armstrong units—suggestive of carcinoma of prostate.

Requirements before estimation of acid phosphatase

- Early morning blood sample
- On empty stomach
- Avoid fatty food
- Per rectal examination should not be done before drawing the sample of blood.

Significance of acid phosphatase

Levels come down with the treatment of carcinoma of prostate, especially when bone metastasis disappears.

4. Serum alkaline phosphatase

It is increased if there is extensive liver metastasis or bone metastasis.

5. Prostate-specific antigen

- Prostate-specific antigen (PSA) is a neutral protease, elaborated by **columnar prostatic acinar epithelial cells**.
- If it is more than 4 nmol/ml, carcinoma is to be suspected; **10 nmol/ml** is suggestive of prostatic carcinoma; **35 nmol/ml:** Disseminated carcinoma.
- The highest concentration of PSA occurs in the lumen of the prostatic acini and ducts (up to million times greater than in systemic circulations). The prostatic luminal cells are normally surrounded by basal cells, prostatic basement membrane and prostatic stroma.
- A number of diseases disrupt some of the barriers to absorption resulting in elevation of serum PSA, **notably prostatic cancer, prostatic inflammation and infarction.** PSA is also transiently elevated (up to 24 hours) after ejaculation and cycling.
- PSA measurement is the most efficient screening test for prostate cancer and it increases further if the measurement is **combined with digital rectal examination** (DRA).
- PSA measurement is also vital in staging prostate cancer and assessing response to treatment.

PEARLS OF WISDOM

Thus, PSA is organ-specific but not cancer-specific.

6. Abdominal and transrectal USG

For staging of the disease

7. CT scan or MRI scan

These are done before proceeding on to radical surgery to assess the extent of the tumour.

8. Bone scan

Is indicated in cases of carcinoma prostate especially who have bony pains, raised alkaline phosphatase and also very high PSA levels (> 20 ng/ml) (*see* clinical notes).

CLINICAL NOTES

A 75-year-old gentleman was admitted to the hospital because of giddiness and syncope. He was a diabetic. Initially, it was thought to be due to hypoglycaemia. Investigations revealed creatinine of 7.5 mg%. Diagnosis of renal failure was made. Urgent ultrasound revealed bilateral hydronephrosis due to large para-aortic nodes compressing both ureters. He underwent emergency stenting of both ureters. Rectal examination revealed a hard prostate. PSA was 20 nmol/ml suggestive of dissemination. CT scan done later showed involvement of pelvic bones, thoracolumbar vertebrae and iliac bones. It was a case of carcinoma prostate. Retrospective analysis revealed that the patient was having backache since the last 6 months.

TREATMENT OF CARCINOMA OF PROSTATE

(Fig. 41.8)

It can be classified under following headings—early malignancy and late malignancy.

I. Early malignancy

It refers to Tl or T2, N0, M0

A. Early prostatic malignancy with PSA levels less than 20 nmol/ml

- **Radical prostatectomy** is done for T1 and T2 and in men with life expectancy of > 10 years. Metastasis should be excluded by a negative bone scan, chest radiograph and serum PSA < 20 nmol/mL. Radical prostatectomy involves pelvic lymphadenectomy and removal of the prostate, seminal vesicle including the distal urethral sphincter followed by anastomosis of urethra to the bladder neck.
- **Most commonly injured vessel**—dorsal venous complex.
- **Radical radiotherapy** for prostate and pelvic nodes is given postoperatively.

Disadvantages of radical prostatectomy

Impotence and stress incontinence may complicate the surgery.

B. Early prostatic malignancy with PSA is 20 nmol/ml or more and the patient is already beyond 65 to 70 years of age, surgery is not favoured. Radical radiotherapy is given.

II. Late malignancy

It means T3 lesions, involvement of regional nodes or presence of metastasis.

1. **Androgen ablation** in the form of bilateral orchidectomy is done as the tumour is androgen-dependent followed by

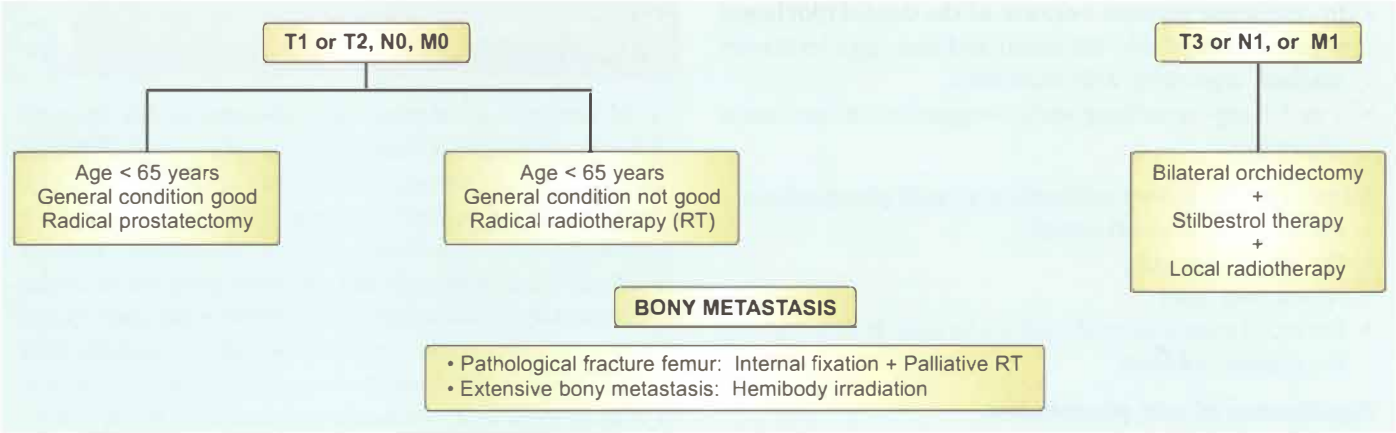


Fig. 41.8: Summary of the treatment of carcinoma prostate

anti-androgenic measures. **Anti-androgenic measures are given below.**

A. Low orchidectomy

Subcapsular orchidectomy used to be done earlier wherein tunica albuginea was left behind. **It is now obsolete because it has been proved beyond doubt that some testicular tissue can still be left behind. Hence, a low bilateral orchidectomy is done.**

B. Drugs—(LHRH agonists, anti-androgens)

1. **Oral stilboestrol** therapy, dose 20–25 mg/day; phosphorylated diethylstilboestrol (Honvon) IV initially later, orally. Within 48 hours of treatment, dysuria improves, pain disappears, bone pains improve and metastasis may disappear.

   Disadvantages
   - Thromboembolism
   - Nausea and vomiting
   - **Gynaecomastia:** Can be prevented by preoperative radiation to the breast—800 rads.

2. **Phosphorylated diethylstilboestrol** is an excellent drug which can be given orally and IV, and the drug is not broken down until it reaches prostate. Hence, systemic toxicity of stilboestrol does not occur. In the prostate it is broken down by acid phosphatase to release stilbestrol locally.

2. Radiotherapy

A. For **localised metastasis** in bones or brain, **radiotherapy** relieves the symptoms.

B. For **generalised metastasis, hemibody irradiation** is excellent to improve the symptoms and the survival time.

C. If carcinoma of the prostate is diagnosed only by a biopsy of the prostate removed during TURP, no surgery needs to be done. **Local radiotherapy** to the prostatic bed is sufficient.

D. If the diagnosis of carcinoma of prostate is made on rectal findings, **surgeons have no role to play** because many a time it is an advanced cancer and there is a danger of bleeding from prostatic venous plexus. Hence, radiotherapy is administered by using linear accelerator (cobalt).

3. Palliative treatment of carcinoma prostate

A. Local radiotherapy to the enlarged para-aortic nodes and to the bone.

B. **Chemotherapeutic drugs**
- Mitomycin and nitrogen mustard are being tried.
- Docetaxel—new drug with better results.

Recent advances in carcinoma prostate

Prostate cancers exhibit heterogenecity within tissue, so **two** histological areas of prostate are each scored between 1 and 5.

The two scores are added to give an overall Gleason score of between **2 and 10**. The score correlates well with the likelihood of spread and prognosis.

Gleason score
- 6 or lower—often candidates for an active surveillance program (previously referred to as watchful waiting therapy).
- 7—usually associated with a critical decision-making step (usually some form of definitive therapy).
- 8–10—often candidates for adjuvant therapy or radiation treatment.

PROSTATITIS

- Inflammation of prostate can be acute or chronic.
- However, in both types, in addition to the prostate, seminal vesicles and posterior urethra are involved.
- Often the diagnosis is delayed because of varying symptoms which will be attributed to a different cause and they are being wrongly treated.

- If the treatment is not effectively given, infection persists and later it is difficult to eradicate.

ACUTE PROSTATITIS

Aetiology

- Causative organisms are *Escherichia coli, Staphylococcus aureus* and *Staphylococcus albus*.
- The infection is usually due to haematogenous spread from a distant focus or secondary to urinary tract infection.
- Instrumentation or invasive urological procedures are also the factors.

Clinical features

- The patient is ill with high grade fever, chills and rigors.
- Pain all over the body, more so in the back.
- Perineal heaviness or pain, rectal irritation and urethral discharge are also the features.
- Pain on micturition is common and initial samples of urine contain 'threads'.
- **Rectal examination finding:** Tender, boggy enlarged prostate. Fluctuation indicates prostatic abscess (rare).

Treatment

- Hospitalisation, intravenous fluids, antipyretics.
- Antibiotics such as ciprofloxacin or norfloxacin should be given for 2–3 weeks. Otherwise, recurrent attacks of prostatitis can occur.
- If **abscess is suspected,** diagnosis can be confirmed by transrectal ultrasound, it can be drained by **transurethral unroofing of abscess** cavity similar in technique to transurethral resection of prostate (TURP).

CHRONIC PROSTATITIS

Chronic prostatitis results due to inadequately treated acute prostatitis (Key Box 41.5).

Clinical features

- Elderly men are affected and they complain of perineal heaviness, perineal discomfort or pain on sexual intercourse.
- Intermittent fever is also a feature
- Rectal examination may reveal a boggy and tender prostate.
- Low backache

Diagnosis

Prostatic massage is given by bidigital method—index finger in the rectum and the thumb in the perineum to one side. Now the patient is asked to void the urine. Presence of prostatic threads or mucopus in the postprostatic massage urine is diagnostic of chronic prostatitis.

Treatment

Prolonged antibiotic therapy—norfloxacin, trimethoprim and metronidazole are used.

KEY BOX 41.5

CHRONIC PROSTATITIS

- Follows recurrent prostatitis
- Elderly men are affected
- Symptoms—misleading
- Perineal heaviness, back pain
- Postprostatic massage—threads in urine
- Pus cells and bacteria—prostatic fluid
- Prolonged antibiotics

INTERESTING WISDOM LINES

- Do not injure prostatic venous plexus (between anatomical capsule and pelvic peritoneum) during prostatectomy or abdominoperineal resection (APR) done for carcinoma rectum.
- Frequency of micturition is not an indication for prostatectomy.
- Most commonly done surgery for BPH is transurethral resection.
- Prostatectomy for BPH does not give protection from development of prostatic carcinoma.
- Androgen ablation is the chief form of treatment in advanced prostatic cancers.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All the topics have been updated.
- New photographs and key boxes have been added.
- Gleason score is added

MULTIPLE CHOICE QUESTIONS

1. Following are true for surgical anatomy of the prostate *except*:
A. Developmentally it has 5 lobes
B. Median lobe enlargement causes carcinoma prostate
C. Develops around 12th week of intrauterine life
D. Between prostate and rectum, Denonvillier's fascia exists

2. Following are the changes in the urinary bladder due to benign prostatic hypertrophy *except*:
A. Facsiculations B. Sacculations
C. Diverticuli D. Carcinoma

3. Vesical introversion of the sensitive prostatic urethra within urinary bladder causes:
A. Frequency B. Urgency
C. Hesitancy D. Haematuria

4. Following are complications of benign prostatic hypertrophy *except*:
A. Stones
B. Renal failure
C. Recurrent urinary tract infection
D. Carcinoma urinary bladder

5. Which of the following drug is used to treat benign prostatic hypertrophy?
A. α adrenergic blockers
B. β adrenergic blockers
C. Oral stilboestrol
D. Oral prednisolone

6. Following are true for role of ultrasound in benign prostatic hypertrophy *except*:
A. Can assess the size
B. Can assess the weight
C. Can assess residual urine
D. Can assess the flow rate

7. Following are true for carcinoma prostate in digital rectal examination *except*:
A. Hard nodule is felt
B. Median sulcus is obliterated
C. Rectal mucosa cannot be moved
D. Ulcerated mucosa

8. The reason for early bone spread from carcinoma prostate to bones is spread through:
A. Batson's plexus
B. Santorini plexus of veins
C. Waldeyer's plexus of veins
D. Denonvillier's plexus of veins

9. Following are the features of bone secondaries fro carcinoma prostate *except*:
A. Thoracolumbar vertebrae are involved
B. Moth-eaten appearance in X-ray
C. Osteolytic
D. Multiple bones are affected

10. Which structure is affected late in carcinoma prostate
A. Prostatic urethra B. Seminal vesicles
C. Rectum D. Urinary bladder

11. Which of the following is not true for aci phosphatase?
A. It is responsible for acidic pH in the prostatic urethr
B. High values suggest carcinoma prostate
C. It is elevated not only in carcinoma prostate but als other conditions
D. Early morning urine sample is the best to measure thi

12. Following are true for prostate specific antigen *except*
A. Released from columnar prostatic acinar epithelia cells
B. More than 4 nmol/ml suggest carcinoma prostate
C. Prostatitis also can increase in the levels
D. It does not help in assessing the response to treatmen

13. Which of the following is not done in cases o carcinoma prostate?
A. Radical prostatectomy
B. Radical radiotherapy
C. High orchidectomy
D. Stilboestrol therapy

14. What is the treatment to follow after prostatectomy done for benign prostatic hyperplasia is reported as carcinoma prostate?
A. Orchidectomy B. Stilboestrol
C. Bisphosphonates D. Local radiotherapy

15. Following are true for acute prostatitis *except*:
A. Usually it is haematogenous infection
B. Instrumentation can also cause this
C. Usually it is mild self-limiting
D. Urine sample may contain 'threads'

16. Which of the following is true for posterior urethral valves?
A. They are acquired
B. Bladder is thin walled and more prone for rupture
C. Renal failure is uncommon
D. They are symmetrical valves

ANSWERS

1 B	2 D	3 B	4 D	5 A	6 D	7 D	8 A	9 C	10 C
11 D	12 D	13 D	14 D	15 D	16 B				

42

Penis, Testis and Scrotum

- Surgical anatomy of penis
- Phimosis
- Paraphimosis
- Carcinoma of penis
- Peyronie's disease
- Anatomy of the testis
- Hydrocoele
- Undescended testis
- Ectopic testis
- Varicocoele
- Spermatocoele
- Epididymal cyst
- Torsion testis
- Testicular tumours
- Fournier's gangrene
- Fracture penis
- What is new?/Recent advances

SURGICAL ANATOMY OF PENIS

- It consists of two corpora cavernosa and one corpus spongiosum (Fig. 42.1).
- **Corpora cavernosa** are vascular spaces wherein **arterioles open directly**. They are corkscrew shaped (**helicine arteries**) which allow their elongation in erection.
- **Corpora spongiosum** is perforated by urethra and is continuous distally with the glans.
- Each corpus is enclosed by tough fibrous membrane—the **tunica albuginea** of the corpus.
- The fused fibrous sheaths are attached to the under surface of the symphysis pubis by a triangular sheet of fibrous tissue called as **suspensory ligament**. It has to be divided during total amputation of the penis.

Blood supply

- The artery to the bulb supplies corpus spongiosum and the glans.
- Deep artery supplies corpus cavernosum alone—its sole function is erection.
- Dorsal artery supplies skin, fascia and glans
- Superficial dorsal vein drains into superficial external pudendal veins. Deep dorsal vein enters prostatic venous plexus.

Penile urethra

- Entire penile urethra is lined with transitional epithelium except dilated anterior part in the glans—fossa navicularis—lined by stratified squamous epithelium.

Lymphatic drainage

- Medial group of superficial inguinal nodes.
- Some from glans pass directly to the node of Cloquet

Nerve supply

- Parasympathetic nerves—nervi erigentes: stimuli resulting in erection of the external genitalia.
- Sympathetic system—hypogastric nerves: helps in ejaculation.

PHIMOSIS

Inability to retract the prepucial skin is described as phimosis.

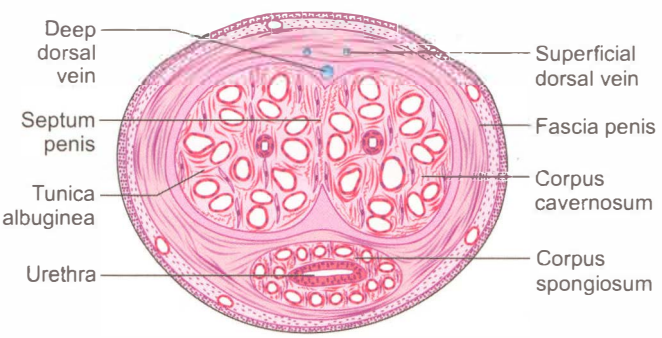


Fig. 42.1: Surgical anatomy of the penis

Causes

1. **Congenital:** Most common type seen in young patients.
2. Secondary to **chronic balanoposthitis:** Balanitis means inflammation of glans penis and posthitis means inflammation of the prepuce. Balanoposthitis is common in diabetic patients.
3. **Chancre** also can cause phimosis.
4. **Carcinoma** of the penis can present as a recent phimosis.

Clinical features

1. Inability to retract the prepuce.
2. In children, ballooning of the prepuce (second bladder) can be seen, which is diagnostic of phimosis.
3. Because of the phimosis, they are more prone for balanoposthitis because of inability to clean the glans.

Complications

1. Carcinoma of the penis (Fig. 42.2)
2. Paraphimosis

Treatment

Circumcision: Removal of the prepuce

PARAPHIMOSIS

In this condition, the retracted skin of the glans penis (prepuce) cannot be pulled forwards. As a result of this, the retracted skin acts like a tight constricting agent which compresses the corona resulting in venous congestion. As venous congestion increases, glans swells up resulting in paraphimosis.

Causes (Key Box 42.1)

1. During catheterisation, if the retracted prepuce is not pulled forwards, it results in paraphimosis.
2. This can follow after a sexual intercourse.

Clinical features

1. Severe pain in the glans penis
2. Gross swelling of retracted prepucial skin and oedema of the distal glans penis.

KEY BOX 42.1

PHIMOSIS—CAUSES

- Cancer
- Chancre
- Congenital
- Chronic disease: Diabetes mellitus

Treatment

1. Sedation
2. Injection hyaluronidase 250 units in 10–15 ml of saline i injected into the constriction ring (prepucial skin which i retracted) 5–10 minutes later, when oedema is reduced, witl gentle manipulation it is possible to reduce th paraphimosis.
3. Dorsal slit is given so that reduction can be done Circumcision follows later.

Complications

1. Ulcers of glans
2. Gangrene of glans in later stages

CARCINOMA OF PENIS

Most common type — squamous cell carcinoma.Othei varieties are—melanoma, adenocarcinoma, basal cell carcinoma (BCC)

Premalignant lesions

1. **Genital warts: Buschke–Lowenstein** tumour is a giant penile condyloma and resembles squamous cell carcinoma. It is a cauliflower-like lesion, may have foci of malignancy.
2. **Erythroplasia of Queyrat** or Paget's disease of penis: a persistent red, raw lesion which is a precancerous condition.
3. **Leukoplakia:** Persistent nonspecific patch in the glans or in the prepucial skin. Interestingly, leukoplakic patches are not white in the penis.
4. **Bowen's disease:** This presents as a small eczematous plaque. Carcinoma *in situ* is a complication which develops within this plaque.

Fig. 42.2: Ca penis proliferative growth. Patient had phimosis

Fig. 42.3: This patient presented with fungating inguinal lymph nodes. Observe the penis—he has undergone partial amputation of the penis 2 years back.

Fig. 42.4: This patient had a large foul smelling proliferative lesion in the glans penis with fungating inguinal lymph nodes. Such cases are advanced cases with poor prognosis

Fig. 42.5: Carcinoma penis difficult to find penis—ask the patient where he passes urine—he will show you the urethral opening

Fig. 42.6: Carcinoma penis—proliferative lesion with gross oedema due to secondary infection

AETIOLOGY

Phimosis

1. Extremely rare in Jews who practise circumcision immediately after birth.
2. Carcinoma penis is rare in Mohammedans who **practise** circumcision few years after birth.
3. Carcinoma penis is common in Hindu and Christian population who do not practise circumcision. Due to the prepucial skin, smegma collects within, which is responsible for chronic irritation giving rise to carcinoma of penis.

PEARLS OF WISDOM

Circumcision done within 1 year confers immunity against Ca penis.

Clinical features

1. Carcinoma penis is common in 6th decade. Majority of patients present with nonhealing ulcer.
2. Foul smelling discharge is common and occasionally it is blood stained (Figs 42.3 to 42.6).
3. Recent phimosis due to growth underneath the prepuce.
4. Haematuria, pain while passing urine indicate locally advanced tumours.
5. On examination, very often there is an ulceroproliferative growth with everted edges and induration of the base and edge. The induration is much more extensive than the lesion. Hence, entire shaft has to be examined for evidence of induration.
6. Urethra is rarely involved in carcinoma of penis because it is protected by the tough Buck's fascia, which is a part of pelvic fascia. In large fungating lesions, it may be

difficult to identify the external urinary meatus. In such situations the patient will point out at the external urinary meatus.

SPREAD

1. Direct spread: Involves the glans, prepucial skin and shaft.
2. Lymphatic spread: Inguinal nodes are enlarged. 30% cases are due to infection. Nodes are firm and tender in infection. Hard nodes suggest metastases. Later, internal iliac and para-aortic nodes can also get enlarged. In advanced cases the lymph nodes may show fungation as in Fig. 42.4.

Cause of death

Cause of death in carcinoma penis: Erosion of femoral vessels by inguinal lymph nodes.

STAGING

I. Tumour confined to the glans or prepuce
II. Tumour involving the penile shaft or corpora cavernosa
III. Mobile regional nodal metastases, with stage I or II
IV. Tumour beyond penile shaft, fixed regional lymph node or distant metastases.

INVESTIGATIONS

1. Wedge biopsy from the edge of the growth, proves the diagnosis of squamous cell carcinoma nodes.
2. FNAC of enlarged inguinal lymph nodes.
3. Ultrasound and MRI are helpful in lesions invading corpora cavernosa (soft tissue details). Ultrasound guided FNAC of lymph nodes.
4. CT scan is useful in obese patients wherein clinical examination of inguinal nodes is difficult
5. CT guided FNAC of pelvic nodes

DIFFERENTIAL DIAGNOSIS

1. Condyloma acuminatum
2. Buschke-Lowenstein tumour
3. Balanitis xerotica obliterans

TREATMENT

It can be divided into treatment of the primary and treatmer of the secondaries.

I. Treatment of the primary

Stage I

1. Growth confined to the prepuce—**circumcision**. Regula follow up is necessary.
2. Growth involving the glans: **Partial amputation** with a least 2 cm margin from the palpable indurated edge of th tumour.

Stage II

1. **Partial amputation:** After having a macroscopic tumour free, 2 cm margin proximally, if there is adequate length o penile shaft (minimum 2.5 cm) to carry out the sexua function and for directing the urinary stream, a partia amputation can be done (Figs 42.7 and 42.8).
2. **Total penectomy with perineal urethrostomy,** if adequate shaft cannot be retained. This is a major operation, patien has to be clearly instructed about the consequences anc complications of perineal urethrostomy.

Complications of perineal urethrostomy

1. **Ammoniacal dermatitis** of scrotum. To prevent this, patient has to lift the scrotum to pass urine.
2. **Stricture of perineal urethra** which can be dilated by Hegar's dilators.

Stage III

- Circumcision, partial amputation or total penectomy followed by ilioinguinal block dissection.

Fig. 42.7: Partial amputation in progress—veins are exposed

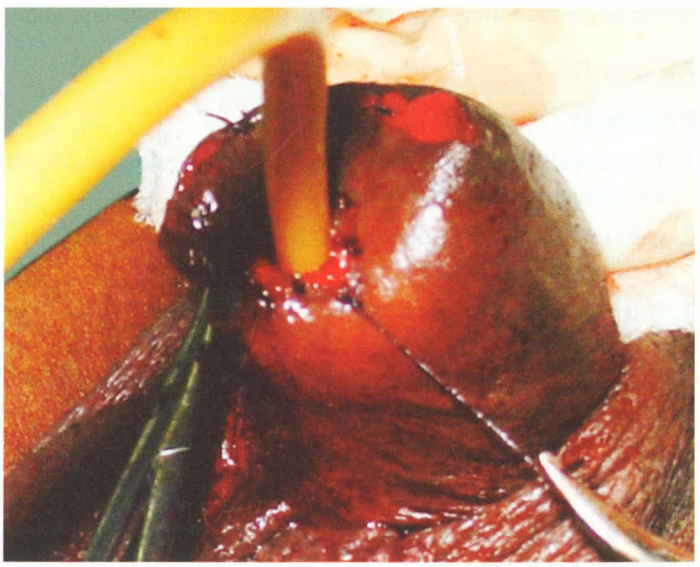


Fig. 42.8: Stump seen with catheter in place

II. Treatment of inguinal lymph node secondaries

- Before discussing ilioinguinal block dissection, it is necessary to know the concept of **sentinel lymph node biopsy (SLNB).**
- The concept of SLNB was first described by Cabana in 1977.
- He demonstrated consistent drainage of the penile lymphatics into a sentinel lymph node or groups of lymph nodes, **located superomedial to the junction of saphenous and femoral veins in the area of superficial epigastric vein.** He postulated that this sentinel lymph node is the first to get involved in the penile malignancy and if this sentinel lymph node is negative for tumour, metastasis to other inguinal lymph nodes will not occur. Metastasis to this lymph node will indicate the need for complete ilioinguinal block dissection.
- **Technique:** A dye, isosulphan blue, is injected at the site of primary tumour. After sometime, the inguinal dissection is done to expose the sentinel lymph node area and the lymph nodes, which take up the dye. These are removed and sent for pathological examination. Based on the report, if node is positive for malignancy, a complete inguinal block dissection is indicated. In case of negative nodes, nothing else is required and patient is kept under regular follow-up.
- Only 50% of patients presenting with palpable lymph nodes actually have metastatic disease, the remainder have lymph node enlargement secondary to inflammation. So, subjecting all the patients with inguinal lymphadenopathy to surgery is not recommended. Hence, a course of antibiotic is given and wait for a period of 4–6 weeks, if the nodes are still palpable, block dissection is carried out.
- However this antibiotic policy can be followed in low grade tumour such as *in situ* carcinoma and T1 lesions. If the lymph nodes regress, wait and watch. If the grade of the tumour is high, do not give antibiotics. FNAC is done and treat accordingly.
- Also, if the nodes are not palpable, and if primary tumour is poorly differentiated, superficial lymph node dissection is done. If the nodes are positive, then modified inguinal block dissection (Catalona) can be done.
- Algorithm (Fig. 42.9) to help in the management of these patients is given below.

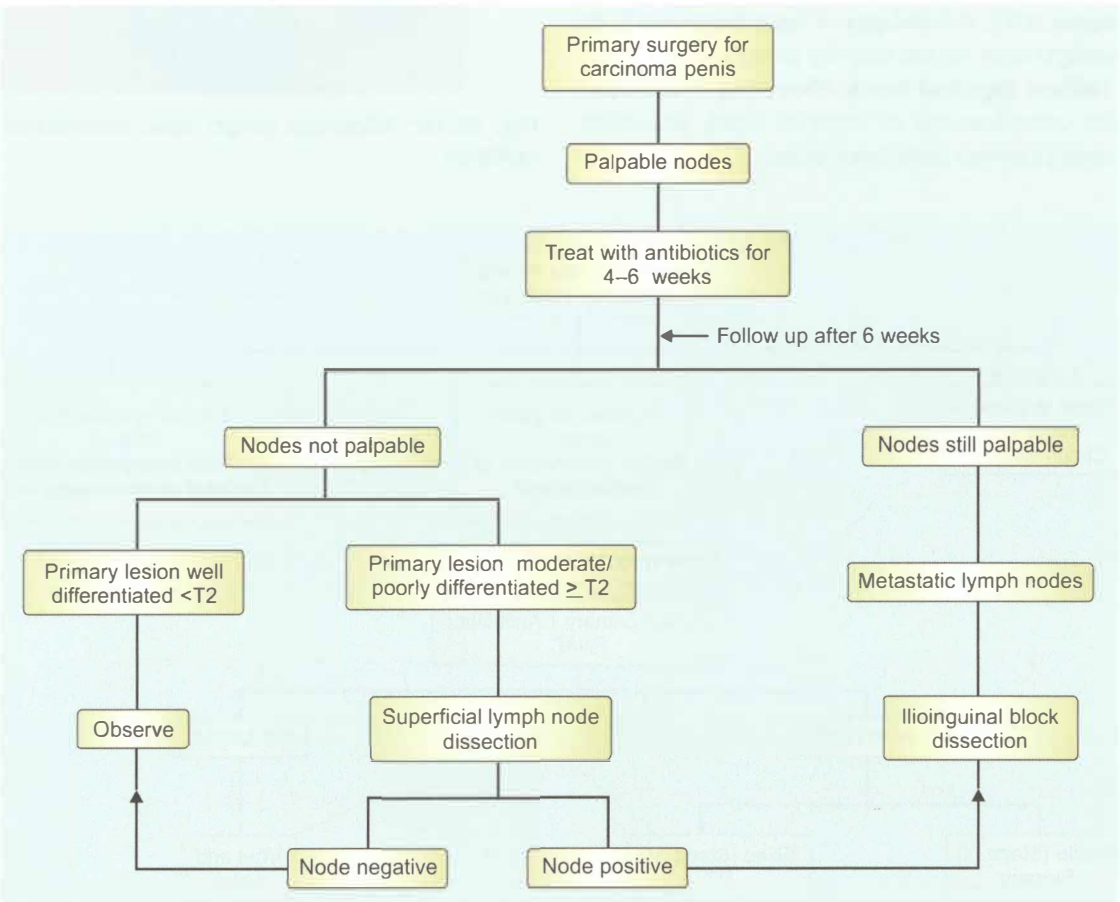


Fig. 42.9: Algorithm of management of inguinal lymph node secondaries from carcinoma penis

PEARLS OF WISDOM

Four weeks of antibiotics are advised after treatment of primary to bring down the infective complications. It does not direct the future management of nodal disease

RECENT ADVANCES IN THE TREATMENT OF CARCINOMA PENIS

ORGAN PRESERVATION

Primary tumours –Tis,Ta,T1; grade 1 and grade 2 tumours – They have favourable histology

Approaches to organ preservation

1. Topical ointments such as 5-fluorouracil or imiquimod cream in Tis only
2. Radiation therapy
3. Mohs surgery: Layer by layer complete excision of penile lesion in multiple sessions with microscopic examination of the undersurface of each layer.
4. Limited excision: It can be done in selected patients who have discrete tumours, with well differentiated carcinoma and after doing an intraoperative frozen section
5. Laser ablation: It is performed in selected patients in conjunction with frozen section biopsies. CO_2 laser has been used widely but recurrence rates are higher – about 40–50% for T1 tumours. Treatment with Nd:YAG laser has least recurrence rates. Advantages of laser treatment is the rates of resumption of sexual activity is high
6. **Modified radical inguinal block dissection (Catalona):** To minimise complications of inguinal block dissection, following modifications have been done.
   - Good preoperative and postoperative care
   - Myocutaneous flap cover
   - Preservation of dermis
   - Preservation of Scarpa fascia
   - Preservation of saphenous vein

Stage IV

Radiotherapy + chemotherapy (cisplatin, methotrexate and bleomycin are the drugs used commonly).

ROLE OF RADIOTHERAPY (Fig. 42.10)

Indications

1. Young patients who want to have a sexual life
2. Patient refuses surgery
3. Fixed/ulcerated inguinal metastasis

Fig. 42.10: Advanced lymph node secondaries treated with radiation

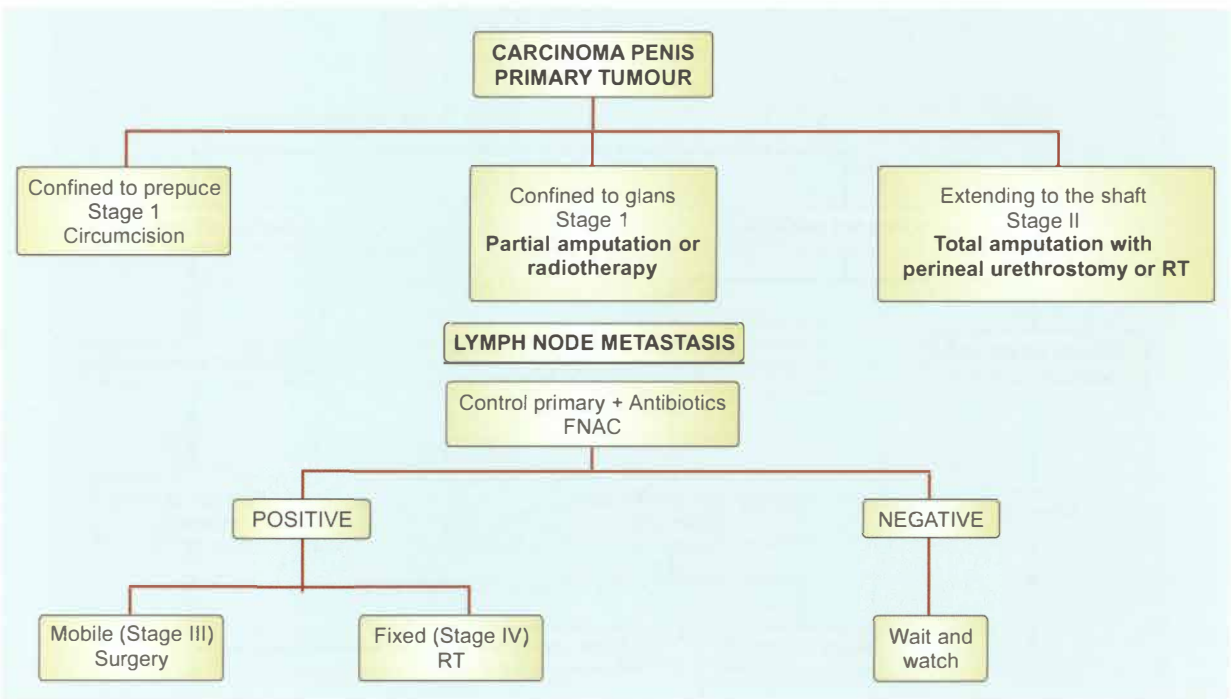


Fig. 42.11: Summary of the treatment of carcinoma of penis

Types

1. External radiotherapy

1. **Dose:** 4000–6000 cGy which can include iliac and inguinal nodes also.
2. **Interstitial radiotherapy:** Iridium wires/tantalum wires are implanted within the glans.

Complication of radiotherapy

- Radionecrosis of penis
- Summary of the treatment (Fig. 42.11).

PEYRONIE'S DISEASE (PENILE FIBROMATOSIS)

Aetiology

- Past trauma has been considered as one of the factors.
- Venereal diseases also have been blamed.
- Association with Dupuytren's contracture, retroperitoneal fibrosis and plantar fasciitis.

Clinical features

- Hard plaques of fibrosis can be palpated along the length of the penis in the sheath of corpora cavernosa (induration – penis – plastica).
- As a result of hard plaques, erection is not proper, and erected penis tends to bend towards the side of the plaque.

Treatment

Medical: Steroids, vit E, tamoxifen (not good results) colchicine therapy, intralesional verapamil

Watch and observe some cases. It may recur after a few years.

Surgical: Straightening of the penis is recommended if the deformity is distressing.

1. **Nesbitt's operation:** Straightening the penis by placing nonabsorable suture in corpora cavernosum opposite the plaque.
2. **Gelhard's operation:** Multiple incisions over fibrous plaques and temporal fascia bridging.

DIFFERENTIAL DIAGNOSIS OF ULCER PENIS

- There are many causes of ulcer penis.
- The important ones being carcinoma and sexually transmitted diseases (Figs 42.12 to 42.14).
- The incubation period is an important clue followed by the nature of the ulcer for the diagnosis.
- Differential diagnosis is given in Table 42.1.

Table 42.1 Differential diagnosis of ulcer penis

	Nature of the ulcer	Inguinal region	Other features/findings
1. Hunterian chancre (syphilitic chancre, hard chancre)	Single, round, painless ulcer-coronal sulcus, frenulum, glans are the sites; base is indurated	Multiple, shotty, painless inguinal lymph nodes	Incubation period—3 weeks; organism. *Treponema pallidum*
2. Chancroid (soft sore)	Multiple painful ulcers; oedematous edges, slough and discharge—plenty of bubo–sinuses	Multiples nodes—above and below inguinal region—**bubo**; suppuration	Incubation period 3–4 days; organism—Ducrey's bacilli
3. Lymphogranuloma venereum (LGV) (lympho-granuloma inguinale, tropical tubular bubo)	Painless vesicles or papules, fleeting duration, heals spontaneously	Multiple nodes above and below in the inguinal region form **sign of groove,** later give rise to bubo and sinuses	Incubation period 1–2 weeks; virus *Chlamydia trachomatis*; rubbery rectal stricture; can occur in females
4. Granuloma inguinale (granuloma venereum)	Painless vesicle, changes into an ulcer with exuberant granulation tissue; highly contagious ulcer; bleeds but painless	Inguinal region may be involved, but inguinal lymph nodes are not involved unless secondary infection supervenes	Incubation period—10–40 days; Organism—*Donovania granulomatis* (bacilli)
5. Balanoposthitis ulcers	Multiple, painful ulcers, difficulty in retracting prepuce	Lymph node enlargement uncommon	Recurrent balanoposthitis common in diabetic patients
6. Herpes progenitalis	Vesicles and pustules on the prepuce or on the glans	Inguinal lymph nodes are not enlarged	Neuralgic pain and itching occurs before the onset of ulcer
7. Carcinomatous ulcer	Painless, indurated ulcer with everted edges; bleeds on touch	Tender nodes, hard nodes. metastasis	Phimosis is one aetiological factor

Fig. 42.12: Multiple painless ulcers due to chancroid

Fig. 42.13: Bubo

Fig. 42.14: Genital chancre

ANATOMY OF TESTIS AND EPIDIDYMIS

Testis

- **Size:** 4 × 3 × 2.5 cm found one in each scrotal sac.
- **Functional unit is a lobule:** 250 lobules filled with seminiferous tubules. It has following cells and their functions are given below:
  Germ cells —→ Sperm production
  Leydig cells —→ Testosterone production
  Sertoli cells —→ Oestrogen production
- The seminiferous tubules converge to form a rete testis, which is connected to the epididymis through 5–7 efferent ductules.
- Covered by thick inseparable covering of fibrous tissue—**tunica albuginea**.
- The serous space in front and lateral surface of testis is **tunica vaginalis** (Fig. 42.15).
- **Blood supply** is by testicular artery—a branch of aorta. The testis gets additional blood supply from artery of the vas and cremasteric arteries. In case of inguinal surgeries, if testicular artery is accidentally injured testicular vascularity is maintained by these two arteries. Veins form pampiniform plexus in the scrotum.
- **Lymphatics:** Drain *via* para-aortic nodes lying along the side of aorta at the level of origin of the testicular arteries (L2) just above umbilicus.

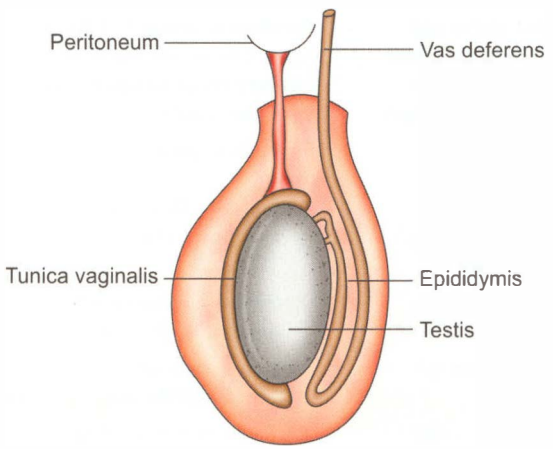


Fig. 42.15: Anatomy of scrotum

Epididymis

- It is 6 m in length (20 feet long), highly coiled and packed and adherent to posterior surface of the testis.
- It has following parts—head, body and tail. Head and body are commonly involved in tuberculosis resulting in posterior sinus formation.
- It is lined by tall columnar epithelium
- The head receives vasa afferentia from the rete testis and is firmly attached to the testis.
- From the tail, the vas deferens (ductus deferens), a direct continuation of the canal of the epididymis, passes up medially.
- Epididymis is supplied by a branch of testicular artery.

HYDROCOELE

Collection of excessive fluid in the tunica vaginalis sac (TV sac).

I. CONGENITAL HYDROCOELE

Occurs due to patent processus vaginalis sac either completely or partially.

Types

1. **Vaginal hydrocoele** (Fig. 42.16)
   Occurs when hydrocoele sac is patent only in the scrotum.
2. **Infantile hydrocoele** (Fig. 42.17)
   The sac from the scrotum is patent up to the deep inguinal ring.

VARIOUS TYPES OF HYDROCOELE

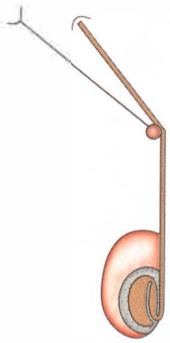


Fig. 42.16

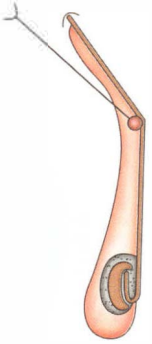


Fig. 42.17

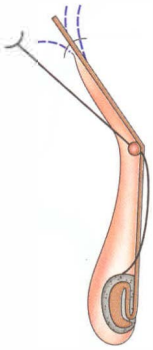


Fig. 42.18

Fig. 42.19

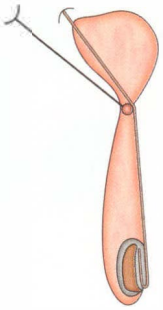


Fig. 42.20

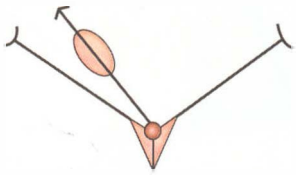


Fig. 42.21

3. **True congenital hydrocoele** (Fig. 42.18)
   - In this condition, the scrotal sac communicates with the peritoneal cavity. It is seen in infants, may be secondary to TB peritonitis. The scrotal swelling appears when the child assumes an erect posture for a long time and it may not reduce due to **inverted ink bottle** effect. Hence, congenital hydrocoele is not reducible. It regresses in size if the child assumes supine position while sleeping.
4. **Encysted hydrocoele of the cord** (Figs 42.19 and 42.23 to 42.25)
   - In this condition the sac is obliterated above (inguinal canal) and below (scrotum) but patent at the root of the scrotum around spermatic cord.
   - It presents as a soft, cystic, fluctuant, transilluminant swelling separate from testis, well above the testis.
   - Diagnosis is established by **traction test**: The swelling has got free mobility but when traction is applied to testis gently, the swelling becomes fixed and it moves down when testis is pulled down. This variety of hydrocoele is treated by excision of the sac.
5. **Hydrocoele-en-Bissac** (bilocular hydrocoele) (Fig. 42.20)
   In this condition, the scrotal sac communicates with another sac underneath the anterior abdominal wall musculature. Diagnosis is made by eliciting **cross-fluctuation** test.
6. **Hydrocoele of canal of Nuck** (Fig. 42.21)
   It presents as a swelling in the inguinal region in female.

II. ACQUIRED HYDROCOELE

a. Primary or idiopathic (Table 42.2 for comparison).
b. Secondary hydrocoele.

PRIMARY VAGINAL HYDROCOELE

This is the most common type of hydrocoele which is seen in young adults, middle age and beyond. It is due to following causes:

1. Defective absorption of fluid
2. Defective lymphatic drainage

PEARLS OF WISDOM

- ***Hydrocoele fluid contains albumin and fibrinogen.***
- ***Filarial hydrocoeles contain liquid fat rich in cholesterol.***
- ***Hydrocoele of hernia is a hernia containing hydrocoele fluid*** (Fig. 42.26)

Clinical features

- Soft, cystic, fluctuant, transillumination positive swelling confined to the scrotum (Fig. 42.22).

Fig. 42.22: Bilateral primary vaginal hydrocoele

Fig. 42.23: Encysted hydrocoele

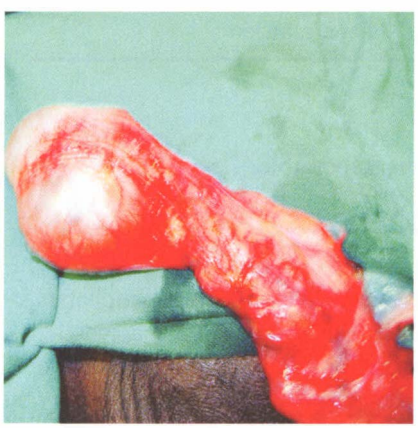


Fig. 42.24: At surgery

Fig. 42.25: Excised specimen

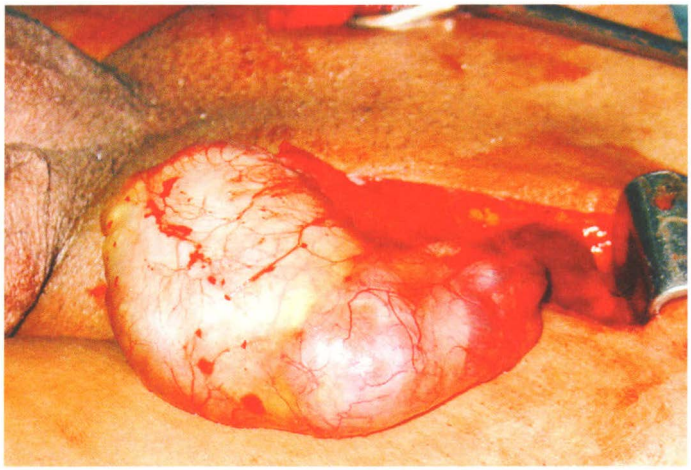


Fig. 42.26: Hydrocoele of hernia

Fig. 42.27: Chylocoele fluid

KEY BOX 42.2

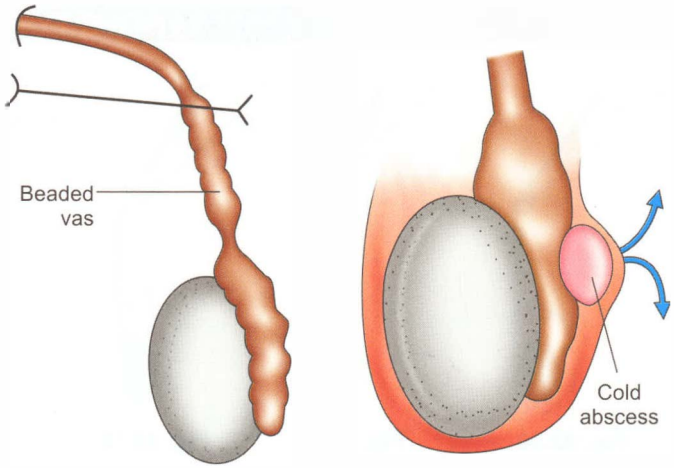
HYDROCOELE: TRANSILLUMINATION NEGATIVE

- Sac is very thick
- Sac is calcified
- Chylocoele (Fig. 42.27)
- Haematocoele
- Pyocoele
- Malignancy testis—blood stained effusion

- Not reducible
- No impulse on cough
- Getting above the swelling is possible.

SECONDARY HYDROCOELE

1. **Recurrent epididymo-orchitis due to filariasis**
   - Fluid that accumulates is due to obstruction of lymphatics. The fluid is milky white. Such hydrocoeles are called chylocoeles and often do not exhibit transillumination. (Key Box 42.2).
2. **Tuberculous epididymo-orchitis** (Figs 42.28 and 42.29)
   - Retrograde infection from the seminal vesicles.



Figs 42.28 and 42.29: Tuberculous epididymo-orchitis

   - **Craggy epididymis** refers to rough, hard, irregular surface. This involves the epididymal head and causes fibrosis. So, the epididymis feels craggy. Vas deferens feels like beads, called as **beaded vas.** Secondary hydrocoele occurs in 30% of the cases. Eventually it forms **cold abscess** which ruptures and results in sinus posteriorly, in the scrotum.
   - It never involves the testis proper.
3. **Testicular tumours:** They can present with a swelling of the scrotum, often diagnosed as hydrocoele. Any young patient with a rapidly growing scrotal swelling could be a testicular neoplasm. Fluid within the sac is haemorrhagic.
4. **Pyocoele:** Infected hydrocoele. Infection in a hydrocoele is rare because of the tunica vaginalis sac which is relatively avascular. However, few cases may get infected resulting in pyocoele. These patients present with fever, chills and rigors.
5. **Haematocoele:** Trauma to the hydrocoele or spontaneous bleeding into the sac.

Comparison of primary and secondary hydrocoele is given in Table 42.2

Treatment of hydrocoele (Key Box 42.3 and also *see* Chapter 53)

1. **Lord's plication** is indicated in small hydrocoeles. The sac is opened and the cut edge of the sac is plicated to tunica albuginea. (It is the reflected portion of the processus

KEY BOX 42.3

TREATMENT OF HYDROCOELE

- Observe
- Lord's plication
- Jaboulay's operation

Table 42.2 Comparison of primary hydrocoele with secondary hydrocoele

	Primary hydrocoele	Secondary hydrocoele
1. Aetiology	Defective absorption of fluid	Excessive production of fluid
2. Examples	Vaginal hydrocoele, infantile hydrocoele.	Filarial hydrocoele, secondary to malignancy of the testis.
3. Size	Moderate, big	Small
4. Palpation of the testis	Difficult	Easily palpable
5. Transillumination	Positive in majority of the cases	Usually negative
6. Consistency	Tensely cystic	Lax, cystic
7. Treatment	Partial excision and eversion	Treatment of the primary

vaginalis.) As a result, the sac gets crumpled up near the testis. The testicular secretions get absorbed by subcutaneous lymphatics and venous system.

2. **Partial excision and eversion of the sac:** Jaboulay's operation. This is indicated in large hydrocoeles. The thick, large, sac is excised and is sutured behind testis.
3. **Aspiration** is a temporary method and there is a chance of introducing infection. It can be done only in high-risk patients. This is a procedure to be condemned.

Complications of hydrocoele

1. Haematocoele: Occurs due to a minor trauma
2. Pyocoele: Infected haematocoele
3. Calcification of hydrocoele sac
4. Rupture of hydrocoele sac—very rare
5. **Hernia of the hydrocoele sac** occurs when there is a small tear in the sac resulting in accumulation of fluid in the subcutaneous planes (Fig. 40.30).

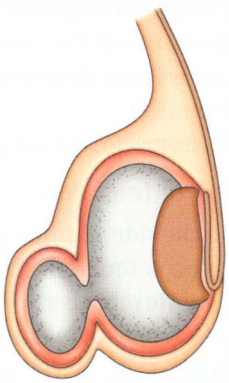


Fig. 42.30: Hernia of hydrocoele sac

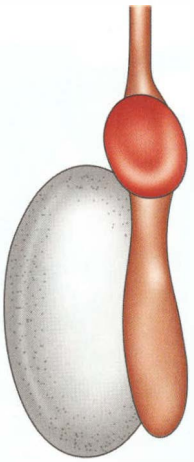


Fig. 42.31: Spermatocoele

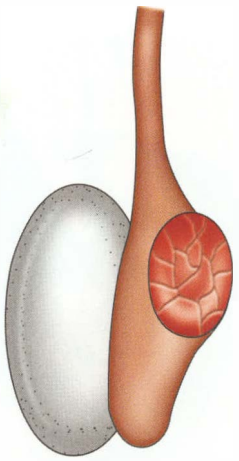


Fig. 42.32: Epididymal cyst

Fig. 42.33: TB sinus scrotum

CYSTIC SWELLINGS IN THE SCROTUM

I. Hydrocoele

II. Retention cyst

1. Spermatocoele (Table 42.3 and Fig. 42.31)
2. Sebaceous cyst (skin of the scrotum) (Figs 42.36 and 42.37)

III. Congenital cyst

1. Epididymal cyst (Fig. 42.32)
2. Cyst of the hydatid of Morgagni

IV. Tubercular epididymo-orchitis

- Cold abscess with a sinus in the posterior aspect of scrotum (Figs 42.33 to 42.35)

Fig. 42.34: Sinus scrotum

Fig. 42.35: Abscess scrotum

Table 42.3 Comparison of epididymal cyst with spermatocoele

	Epididymal cyst	Spermatocoele
1. Aetiology	Cystic degeneration of the appendages of epididymis—**congenital**	Obstruction to the sperm conducting mechanism; **acquired**—retention cyst
2. Site	Behind and above the testis in the region of epididymal head	Behind the body of the testis
3. Loculi	Multilocular	Unilocular
4. Contents	Crystal clear, watery	Barley water-like
5. Transillumination	Brilliant (Chinese lantern pattern)	Poor transillumination—very often negative
6. Aspiration	Results in recurrence as the cyst is multilocular	May cure as the cyst is unilocular
7. Excision	Excision may be necessary if the cyst is large	May be excised if aspiration is not successful

Fig. 42.36: Multiple sebaceous cysts of scrotum (*Courtesy:* Prof Abdul Majeed, MS FRCS, Yenepoya Medical College, Mangalore)

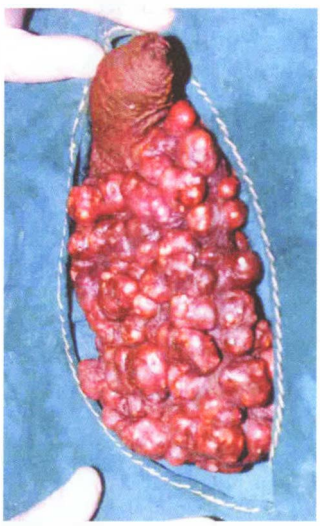


Fig. 42.37: Strawberry scrotum (*Courtesy:* Dr Umesh Bhat, General Surgeon, Kundapur, Karnataka)

UNDESCENDED TESTIS

When the descent of testis is arrested somewhere in its normal pathway to the scrotum, undescended testis occurs.

Development

- The testis develops in the retroperitoneum close to the posterior abdominal wall from the genital tubercles.
- It is guided to the scrotum by the gubernaculum.
- By around 7th month, it reaches the deep inguinal ring, 8th month—inguinal canal and 9th month—superficial inguinal pouch. In normal situations, the testis reaches the scrotum at full term.
- As it comes down, it is surrounded by processus vaginalis sac. This sac gets completely obliterated in normal persons. The persistence of the processes vaginalis sac is responsible for development of a hernia and hydrocoele.

Causes

1. **Muscular hypotonia:** The descent of the testis depends upon muscular contractions of the anterior abdominal wall. Hence undescended testis is seen in children with poor muscle tone, e.g. prune-belly syndrome and Down's syndrome
2. **Gubernaculum dysfunction**
3. **Maternal human chorionic gonadotropin** (HCG) which causes development (maturation) of testis and also helps in descent of the testis. If there is deficiency of HCG, imperfectly developed undescended testis appears.
4. **Familial**
5. Retroperitoneal adhesions prevent the descent of the testis.

Clinical features

1. Right side is more often involved. Bilateral undescended testis is found in about 20% of cases.
2. **Cryptorchidism:** When both testes are impalpable as in cases of abdominal testis and inguinal testis.
3. **Retractile testis:** In this condition the scrotum is well developed, the testis is palpable at the root of the scrotum and can be brought down to the scrotum. Retractile testis is harmless and spontaneously gets corrected within 1–2 years of age, without any treatment. The squatting position may help in such cases, in diagnosing the condition as well as its descent down to the scrotum.

Complications (can be remembered as TESTIS)

T Trauma produces pain
E Epididymo-orchitis will mimic an acute abdomen.
S Sterility: Histological changes start at the age of 2 and by the age of 12, irreversible damage occurs to the spermatogenesis, due to atrophy of testis. Endocrine function will remain normal.
T Torsion
I Indirect hernia in majority of cases.
S Seminoma of testis and other testicular malignancies are reported in greater frequency in undescended testis than in normal testis. **(Risk of seminoma in both testis, not only in undescended testis.)**

Treatment

1. Treatment of choice is orchidopexy.
   - It can be done by open method or laparoscopic method.
   - It can also be a one-stage or two-stage procedure.
   - 2 stage procedure for undescended testis
     (Fowler-Stephens technique)
     Stage I — tethering spermatic blood vessels are divided
     Stage II— testicle placed into scrotum after collateral blood supply to testicle has developed.

   Procedure
   - Considering the psychological, functional and malignant potentials, 6 months to 1 year is the ideal age to operate in bilateral undescended testis and 4 years for unilateral cases. The inguinal canal is explored, testis mobilised by dividing the adhesions and brought down into the scrotum and fixed by using nonabsorbable suture material.
   - A dartos pouch can be formed and followed later by narrowing of the root of scrotum.
   - Associated hernial sac is excised
2. Orchidectomy is done after the age of 14 because of malignant potential.
3. **Ombredanne's procedure:** Testis is brought down to the opposite scrotum through scrotal septum and kept in dartos pouch.
4. **Silbar procedure:** Testicular artery and vein can be divided and reanastomosed using microvascular techniques.

ECTOPIC TESTIS

- Testis is present in an **ectopic** site (not the route through which is descends).
- **Sites** of ectopic sites are:
  - Superficial inguinal pouch
  - Perineum
  - Root of the penis
  - Femoral triangle (thigh)
- Anatomically the size is normal. Physiologically, it functions normally.
- **Embryology:** Testis reaches the scrotum by the scrotal tail gubernaculum. However, if this is weak, the other scrotal tail may pull it in a different direction, resulting in ectopic testis (Lockwood's theory).
- **Complication:** It is more prone for injuries
- **Treatment:** Orchidopexy in a new scrotal pouch

VARICOCOELE

Definition

- Dilatation of testicular veins with/without cremasteric veins which drain into testicular veins.
- Testicular veins which drain the testis and epididymis, form multiple veins in the scrotum which are called as pampiniform plexus of veins. As they travel the inguinal canal, they are reduced to 6–8 in number. At the level of deep ring, they are 2 in number and in retroperitoneum, it forms the single testicular vein.
- On the right side, the testicular vein drains into inferior vena cava (IVC) directly.
- On the left side, it drains into the left renal vein at right angles where there is a valve.

Aetiology

1. Varicocoele is common on the left side (because left testis is at a lower level than the right). The flow of the blood from the left side is into the renal vein where it makes an angle of 90 degrees.
2. Congenital absence of valves.
3. A recent varicocoele in an elderly patient suggests renal cell carcinoma invading the renal veins.

Clinical features

- Common in thin, tall patients
- Hot climates, favour the development of varicocele.
- In the **standing position**, the diseased side appears to be more swollen than the other side. It feels like a bag of worms. On asking the patient to cough, there is fluid thrill, due to regurgitation of venous blood. On the side of varicocoele, scrotum is at a lower level.
- On asking the patient to lie down, it is reducible (disappears).
- Dragging pain in the scrotum is a feature but it is nontender.
- Testis may appear small and soft with diminished testicular sensation.
- **Blow test:** On blowing, fluid thrill may be felt, it may increase in size (Valsalva's manoeuvre).

  An USG demonstrating veins 3.5 mm or more in diameter with reversal of venous flow after Valsalva manoeuvre– varicocoele.

Grading of varicocoele

I (small) — palpable only with Valsalva manoeuvre
II (moderate) — palpable without Valsalva manoeuvre
III (large) — visible through scrotal skin.

Complications

1. Oligospermia
   This occurs due to following reasons:
   A. The **venous congestion** due to the varicocoele results in increased temperature in the scrotum which is supposed to affect spermatogenesis.
   B. **Reflux of the blood** from the renal vein brings powerful hormones secreted from adrenal glands like cortico-steroids and adrenaline which may suppress spermato-genesis.

2. Hydrocoele (most common) — due to ligation of lymphatic vessels.
3. Recurrence.

PEARLS OF WISDOM

Oligospermia is the major but significant complication of varicocoele.

Treatment

1. **Inguinal approach:** Excision of pampiniform plexus in the inguinal canal after ligating them. Testis still has a venous drainage through cremasteric veins.
2. **Retroperitoneal approach** (Palomo's operation): In the retroperitoneum, testicular vein is single and is separate from vas deferens. Hence, it is ligated up in the retroperitoneum. This operation was once thought better than inguinal approach since there is no danger of damaging the vas and ligation of testicular vein is easy but recurrence rates are high and hence not favoured nowadays.
3. **Subinguinal microscopic** varicocelectomy for complete ligation.

TORSION TESTIS (TORSION OF SPERMATIC CORD)

Predisposing causes

- **Inversion of testis** is the commonest cause where testis lies horizontally or upside down.
- **High investment** of tunica vaginalis—the Bell Clapper deformity.
- In cases where the body of testis is separated from the epididymis.
- **Sudden contraction** of spirally attached cremasteric muscle leads to rotation of testis around the vertical axis during straining at stools, lifting heavy weight, coitus.
- **A long redundant spermatic cord** allows twisting of the testis on its own axis.

Two types of testicular torsion:

- **Extravaginal torsion:** It is diagnosed in newborns and is caused by non-adherence of tunica vaginalis to the dartos layer. As a result, spermatic cord and tunica vaginalis are rotated as a unit.
- **Intravaginal torsion:** It is usually diagnosed in boys of 12–18 years of age, but it can occur at any age.

Clinical features

- Age: 10–25 years, sudden agonising pain in the groin and lower abdomen and may be with vomiting.
- **Scrotum is empty** and oedematous on the side of lesion.
- Tender lump at the external abdominal ring—the testis is positioned high **(Deming's sign)**
- **Prehn's sign:** Elevation of scrotum increases pain in torsion of completely descended testis (decreases pain in epididymo-orchitis)
- **Angell's sign:** The opposite testis lies horizontally because of the presence of mesorchium.

Management

- Best results—if detorsion occurs within 4 hours of onset of pain.
- **Scrotal doppler** to confirm the diagnosis
  1. In the first hour untwist the testis manually
  2. If this is not successful urgent exploration of the scrotum and undo the torsion and viable testis should be fixed to the scrotum to prevent recurrence
  3. Gangrenous testis should be removed
  4. Opposite side testis should be fixed at an early date to prevent torsion as it also has higher risk of undergoing torsion.

PEARLS OF WISDOM

Patients should be counselled and consented for orchidectomy before exploration.

TESTICULAR TUMOURS

They constitute 1% of all malignant tumours in the males and almost all are malignant (more than 99%).

CLASSIFICATION

A. WHO classification (Key Box 42.4)

KEY BOX 42.4

WHO CLASSIFICATION GERM CELL TUMOUR

Seminoma	Nonseminomatous germ cell tumours
• Classic (85%)	• Teratoma
• Spermatocytic	• Embryonal cell carcinoma
• Anaplastic	• Choricocarcinoma
• Lymphocytic	• Yolk sac tumour

B. Other classification

I. Seminoma is the most common germ cell tumour: 50% incidence

Types

1. Spermatocytic type—good prognosis
2. Lymphocytic type
3. Anaplastic type

II. Teratoma

- Incidence: 30% (subtype will be discussed later)

III. Combined

- 10 to 20%

IV. Interstitial cell tumours

a. Leydig cell tumour

b. Sertoli cell tumours

V. Lymphoma of testis: Very rare

SEMINOMA

Aetiology

1. **Undescended testis,** undoubtedly predisposes to seminoma.
   - 1 in 20 abdominal testis, 1 in 60 testis at the level of deep ring and 1 in 80 inguinal testis are prone for testicular tumours.
   - However, it should be noted that 25% of testicular cancers in patients with crypto-orchidism occur in normal, descended testis.
2. **Klinefelter's syndrome:** These patients are prone for development of seminoma testis. Other features of the disease are testicular atrophy, absence of spermatogenesis, gynaecomastia, etc.
3. **Trauma** to the testis is a coincidence. This may not precipitate a testicular tumour but draws the attention of the patient to it.

Pathology

- **Seminoma** arises from the seminiferous tubules. As the tumour grows, it compresses the normal testicular tissue. The cut surface is smooth, homogenous (Fig. 42.38). **Microscopy: Round to oval cells** with prominent nucleus. In few cases, **lymphocytic infiltration** can be found.

Types of seminoma

1. **Typical: It is the commonest type of seminoma.** Also called as classic variety. Syncytiotrophoblastic type produces high levels of β-HCG.

Fig. 42.38: Seminoma testis *(Courtesy:* Prof. Sashidharan, KMC, Manipal)

2. **Spermatocytic type**: It occurs in elderly patients. It is slow growing, rarely spreads. Hence has good prognosis.
3. **Anaplastic**: As the name suggests, this variety has high mitotic index/anaplasia, thus spreads fast and carries poor prognosis.
4. **Atypical form**

TERATOMA

Teratoma arises from rete testis. The tumour contains totipotential cells and so can have ectodermal, mesodermal and endodermal elements (Fig. 42.39).

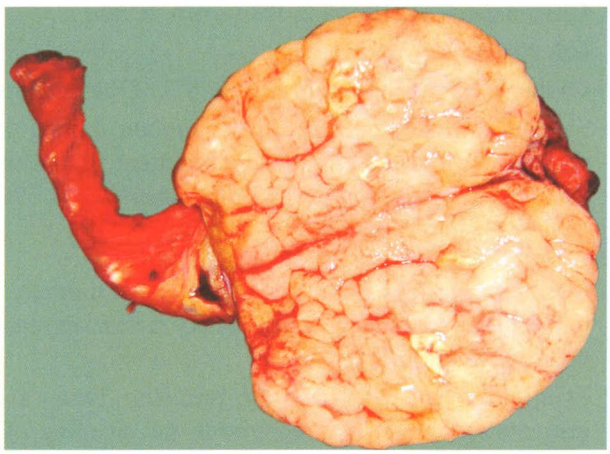


Fig. 42.39: Testicular tumour. Whole of testicular parenchyma is replaced by tumour

Types of teratoma

1. **Malignant teratoma differentiated:** It is the least common variety (1%). It is almost a benign tumour—**Dermoid cyst.** Orchidectomy cures the disease.
2. **Malignant teratoma intermediate:** This is the most common variety (30%) of teratoma containing malignant and incompletely differentiated components.
3. **Malignant teratoma anaplastic:** Highly malignant tumour. Secretes alfa fetoprotein (AFP). Cells are presumed to be from yolk sac.
4. **Malignant teratoma trophoblastic:** This is an uncommon tumour (1%). This secretes very high levels of β-Human chorionic gonadotropins (β-HCG).

CLINICAL FEATURES OF TESTICULAR TUMOURS

(Key Box 42.5)

I. Typical presentation

- **Age:** Teratoma 20–30 years, seminoma 30–40 years (Fig. 42.38)
- **Testicular swelling:** More often heaviness rather than hypertrophy or if it is infiltrated with tumour but vas is never involved. This is called **sign of vas negative** (sign

KEY BOX 42.5

TESTICULAR CANCER

- Irregular testis
- Indurated testis
- Nodular testis
- Nontender enlarged testis
- Young age testicular mass, small hydrocoele
- Heaviness, loss of sensation

of vas positive in TB epididymo-orchitis where there is beading of vas).

- **Haemospermia:** Blood in the semen, is rare
- **Infertility:** Not uncommon
- **Gynaecomastia** is seen in about 10% of the patients.
- Secondary hydrocoele is not uncommon. Young adult with small hydrocoele and enlargement of the testis should arise suspicion of testicular caner

II. Atypical presentation

1. Hurricane variety is the most malignant tumour. The tumour grows rapidly with pulmonary metastasis (cannon ball) and death in a few days.
2. Mimicking acute epididymo-orchitis: This variety presents as severe pain along with the swelling of the testis but does not respond to antibiotics.

III. Symptoms mainly due to metastases

1. Lymphatic spread

- Para-aortic node mass—distension of the abdomen (Fig. 42.40).
- Left supraclavicular node mass—swelling in the neck.
- Iliac node mass—swelling of the leg.

2. Blood spread: Extensive pulmonary secondaries occur from a malignant teratoma.

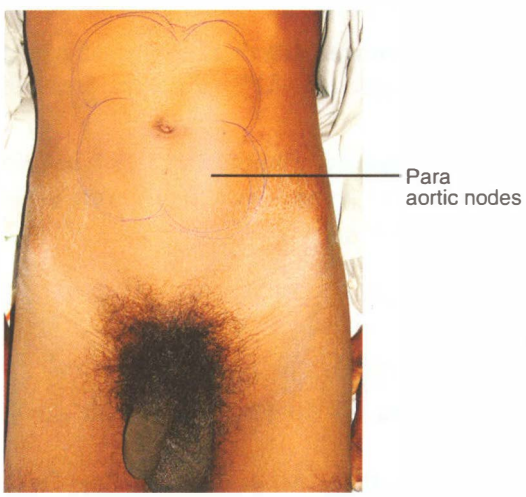


Fig. 42.40: Seminoma testis with para-aortic nodes—clinical examination of testis is very important in an upper abdominal mass

TNM STAGING Testicular carcinoma[1]

T: Primary tumour
Tx: Tumour cannot be assessed
T0: No primary tumour
Tis: Carcinoma *in-situ*
T1: Tumour limited to testis and epididymis without vascular or lymphatic invasion (tumour may invade tunica albuginea but not vaginalis)
T2: Tumour limited to testis and epididymis + vascular or lymphatic invasion or invasion of tunica vaginalis
T3: Tumour invades spermatic cord (with or without vascular or lymphatic invasion)
T4: Tumour invades scrotum with or without vascular or lymphatic invasion
N: Regional lymph nodes
Nx: Lymph node status cannot be assessed
N0: No lymph node metastasis
N1: Single or multiple < 2 cm in greatest dimension
N2: Nodes 2–5 cm in greatest dimension
N3: Nodes > 5 cm in greatest dimension
M: Distant metastasis
Mx: Distant metastasis cannot be assessed
M0: No distant metastasis
M1: Distant metastasis present
M1a: Nonregional nodal or lung metastasis
M1b: Nonpulmonary visceral metastasis (M2)

[1]Staging also considers serum tumour markers -S-LDH, HCG, AFP

SPREAD

- Seminoma spreads by lymphatic route. Along the testicular vessels it spreads to para-aortic lymph node mass, through thoracic duct, to mediastinal nodes and left supraclavicular nodes. ***Spread does not occur to inguinal nodes unless scrotum is incised.***
- Malignant teratomatous tumours spreads predominantly by blood.

STAGING OF TESTICULAR CANCER

- Stage I: Tumour confined to the testis only.
- Stage IIA: Tumour and lymph nodes below the diaphragm—size less than 2 cm.
- Stage IIB: Tumour and lymph nodes below the diaphragm—size more than 2 cm.
- Stage III: Tumour and lymph nodes above the diaphragm.
- Stage IV: Blood spread to lungs/liver/elsewhere.

INVESTIGATIONS

1. **No biopsy** should be done through scrotal route because if the scrotal skin is involved, the spread occurs to inguinal lymph nodes opening up one more channel of lymphatics. Even FNAC is not recommended.
2. **Chest X-ray:** To rule out cannon ball secondaries as in teratoma.

3. **Ultrasound of the testis:** Seminoma appear as hypoechoic lesion and nonseminomatous tumours as inhomogenous.
4. **Abdominal ultrasonography** to see for enlarged lymph nodes, secondaries in the liver, or to detect a tumour in an undescended testis. However, CT scan is a better investigation.
5. **CT scan:** Heterogenously enhancing testicular mass with retroperitoneal lymph nodes and liver metastasis can be made out (Fig. 42.41).
6. **24 hours urine sample for HCG**
   - Normal levels—less than 100 IU
   - More than 1000 IU is diagnostic of choriocarcinoma
   - Hence, it is the tumour marker of choriocarcinoma.
7. **Human chorionic gonadotrophin: Serum HCG** (Key Box 42.6)
   - As the name suggests, it is made by chorionic elements.
   - HCG as a hole (α- and β-HCG) may be increased in testicular neoplasm, melanoma, lymphoma, etc. It can also be raised in nonmalignant conditions such as cirrhosis, peptic ulcer disease.
   - It is the β-HCG which is more important in diagnosing testicular neoplasms and also useful in the postoperative period to know residual tumour or recurrent tumour.
   - The blood levels of **β-HCG is 0 ng/ml.**
8. **α-fetoprotein:** Increased in nonseminomatous germ cell tumours.
9. **Lactate dehydrogenase (LDH):** Increased in nonseminomatous germ cell tumours. LDH levels indicate tumour load. It is not specific for any tumour.
10. **Placental alkaline phosphatase** is increased in seminoma testis.

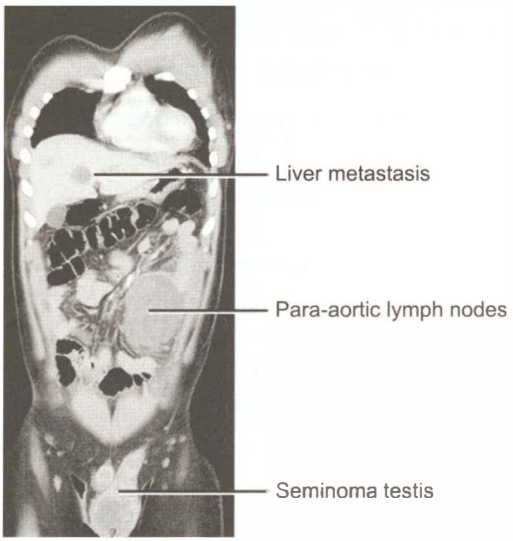


Fig. 42.41: Testicular tumour with para-aortic lymph nodes and liver metastasis

KEY BOX 42.6

β-HCG

- Never found in normal persons
- Secreted by syncytiotrophoblasts
- Choriocarcinoma: 100% cases,Seminoma: 10% of cases
- Embryonal carcinoma: 65% of cases
- Increased levels after orchidectomy indicate recurrence, residual tumour.
- Tumour marker of teratocarcinoma.

TREATMENT

I. Treatment of the tumour

- **Radical inguinal orchidectomy is the treatment of choice in all testicular tumours irrespective of the histological type and stage.**

When a patient presents with rapidly growing testicular swelling and the neoplasm is doubtful, **testis is explored through an inguinal incision.** It is delivered out and a **soft clamp is applied to the testicular vessels at the level of deep ring while doing the procedure so that tumour embolisation does not occur**. Testis is split open, the suspicious area is biopsied and sent for frozen section. If the frozen section is positive, the cord and testicular vessels are divided at the level of deep ring and testis is removed. This is called as **high orchidectomy**. If frozen section is negative, the testis is sutured back and replaced in scrotum. This kind of procedure done through inguinal route is called as **Chevasu's procedure**.

II. Treatment of the retroperitoneum and metastasis

(Key Box 42.7)

1. Seminoma

a. **Stage I-IIA (Low stage):** Radiotherapy to retroperitoneum (2500–3000 cGy) is the treatment of choice:

KEY BOX 42.7

RETROPERITONEAL LYMPH NODE DISSECTION

- It is recommended in all cases of teratoma including stage I. Also indicated in residual disease in lymph nodes in seminomatous tumours also.
- All groups of lymph nodes draining the tumour such as precaval, paracaval, interaortocaval, retroaortic, paraaortic, common iliac nodes are removed.
- It also involves removal of gonadal vein and fibrofatty tissue around the vein from its origin near the internal ring to its insertion to renal vein on the left side and inferior vena cava on the right side.
- The procedure is done on both sides. Haemorrhage, injury to ureter and bowel are other complications.
- Retrograde ejaculation is one common problem after RPLND
- Haemorrhage, bowel and ureter injury are common.

- 5-year survival rate is around 95%.
- Relapse after radiotherapy is managed by chemotherapy.

b Seminoma stage IIB, III, IV

- Radical orchidectomy + chemotherapy-PVB regimen-cisplatin, vincristine, bleomycin.
- BEP: Bleomycin, etoposide and cisplatin

c. Treatment of residual disease in lymph nodes

- In stage IIB and III, if there is residual lymph nodal mass 3 cm in size even after chemotherapy, then, a retroperitoneal lymph node dissection (RPLND) has to be done.
- Stage IIB and III: Survival for 5 years—75%
- Stage IV: Poor survival.

II. Teratoma

a. Stage I, IIA (Low stage)

- Radical orchidectomy and RPLND 5-year survival–85%

b. Stage IIB, III

Radical orchidectomy and chemotherapy (PVB), post chemotherapy residual tumour in the retroperitoneum and if the tumour markers levels regress—retroperitoneal lymph node dissection should be done. 5-year survival rate is around 60%.

c. Stage IV

Orchidectomy + chemotherapy—poor prognosis.

Comparison of seminoma with teratoma (Table 42.4)

Other tumours are given below:

INTERSTITIAL CELL TUMOURS

- 2 types are important clinically.
- Leydig cell tumour and Sertoli cell tumour.
- They should be treated like teratoma.

a. Leydig cell tumour

- It is prepubertal tumour
- Causes masculinisation due to increased androgens—infant Hercules
- Spreads to lymph nodes and lungs
- Good prognosis because it behaves almost like a benign lesion

b. Sertoli cell tumours

- They are feminising tumours: Gynaecomastia, loss of libido, and aspermia are other features.
- Increased oestrogen production is responsible for feminisation.
- Postpubertal tumour
- Orchidectomy is the treatment of choice.

FOURNIER'S GANGRENE (IDIOPATHIC GANGRENE OF SCROTAL SKIN)

Aetiopathology

Even though Fournier's gangrene is called as idiopathic gangrene, certain factors precipitate the scrotal gangrene.

1. Low socio-economic group patients.
2. Unhygienic conditions

Following perianal abscesses, urogenital instrumentation a scratch, cut or bruise in the scrotal skin (instrumentation, injury, infection—Key Box 42.8).

KEY BOX 42.8

FOURNIER'S GANGRENE

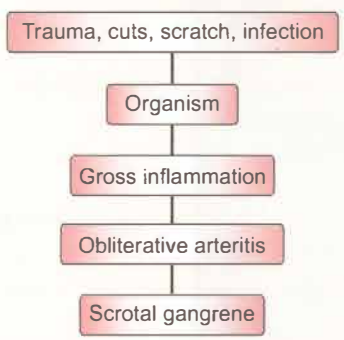


Table 42.4 Comparison of seminoma with teratoma

	Seminoma	Teratoma
1. Cell of origin	Seminiferous tubules in the mediastinum of the testis	Totipotential cells in the rete testis
2. Incidence	35–40%	30%
3. Age group	30–40 years	20–30 years
4. Shape of testis	Retained	Not retained
5. Surface	Smooth	Irregular
6. Cut surface	Smooth	Variegated—nodules, cysts
7. Consistency	Firm	Firm or soft (cystic)
8. Spread	Mainly lymphatic	Predominantly blood
9. Tumour markers	–	HCG—malignant teratoma
10. Radiation response	Excellent—melts like snow	Less sensitive

Fig. 42.42: Fournier's gangrene (*Courtesy:* Prof Santhosh Pai, Manipal)

Fig. 42.43: Fournier's gangrene precipitated by perianal abscess

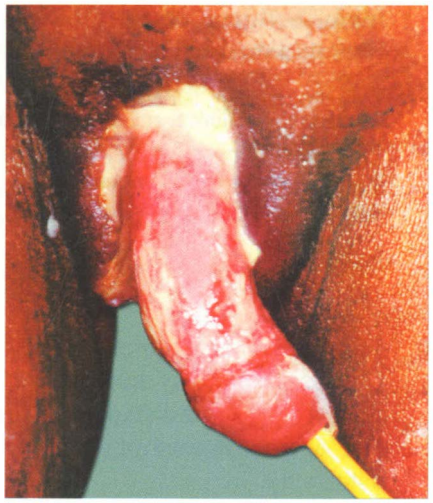


Fig. 42.44: Perineal phlegmon (*Courtesy:* Dr Ramachandra L, KMC, Manipal)

Causative organisms

- Microaerophilic *Haemolytic streptococci*
- *Staphylococci*
- *E. coli*
- Anaerobes: *Clostridium welchii*

(Can be compared to **Meleney's ulcer**—synergistic gangrene—affecting abdominal wall skin. Today it is grouped under necrotising fasciitis).

Clinical features

1. Common in young apparently healthy individuals
2. Sudden appearance of scrotal inflammation—red, swollen, very painful. Patient is toxic with fever, prostration.
3. Within one/two days, extensive gangrene of the scrotal skin occurs resulting in sloughing of the scrotal skin exposing the testicles. In few cases, the gangrene can involve skin of the penis, anterior abdominal wall, medial side of thigh, perianal region. In such situations, it is described as **perineal phlegmon** (Figs 42.42 to 42.44).
4. Luckily, the testis does not get involved in Fournier's gangrene because of thick tunica albuginea.

Treatment

1. Broad spectrum antibiotics are started, once pus is sent for culture and sensitivity, e.g.
   - Metronidazole for anaerobes
   - Gentamicin for gram-negative organisms
   - Ampicillin for gram-positive organisms
   - Cephalosporins may have to be added if required.
2. Gangrenous portion of the scrotum has to be excised, as soon as possible which brings a dramatic reversal of general condition of the patient from toxic to near normal.
3. If the testicles are exposed, they can be implanted in the thigh or once the inflammation subsides, skin grafting is done to cover testicles.

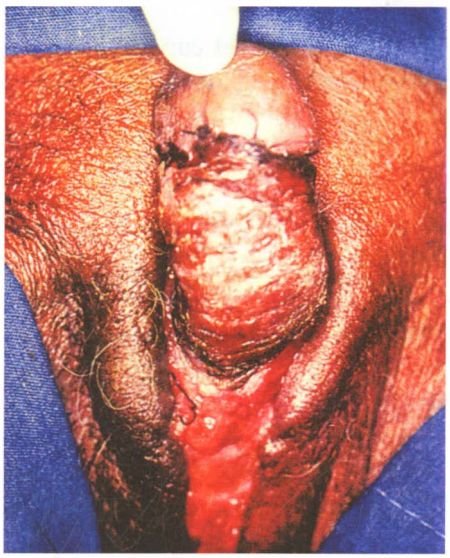


Fig. 42.45: Healing with skin grafting (*Courtesy:* Dr Ramachandra L, KMC, Manipal)

4. If penile skin is gangrenous, it is excised and covered with split skin graft later. Surprisingly, results are better than expected!! (Fig. 42.45)

MISCELLANEOUS

FRACTURE OF PENIS

It is a misnomer

- It is the traumatic rupture of corpora cavernosum. It is considered a urologic emergency.
- Sudden blunt trauma or abrupt lateral bending of penis in an erect state can break the markedly thinned and stiff tunica albuginea.

- One or both corpora may be involved and concomitant urethral (38%) injury if both corpora involved.
- **Causes:** Sexual activity, masturbation, gun shot wounds, industrial accidents, mechanical trauma.
- Diagnosed clinically.
- In equivocal cases — cavernosonography or MRI.
- Preoperative retrograde urethrography urethral injury is suspected.

Treatment

a. Medical: Fluids, antibiotics.
If surgical therapy is delayed due to urethral injury, initial medical therapy consists of cold compresses, pressure dressings, anti-inflammatory medications and suprapublic cystostomy.

b. Surgical: Evacuate haematoma, identify site of injury, correct the defect in tunica albuginea and repair urethral injury urethral catheter removed after 2 weeks.

Complications

Erectile dysfunction, abnormal curvature, painful erections, urethrocutaneous fistula, corpora-urethral fistula and painful nodules.

PRIAPISM

- Priapism is a pathologic condition of a penile erection that persists beyond or is unrelated to sexual stimulation.
- It can occur in all age groups including the newborn but peak incidence:
  5–10 years
  20–50 years.

Causes

1. **Low flow:** Sickle cell trait/disease, chronic myeloid leaukemia, total parenteral nutrition, medications (sildenafil, cocaine), malignant penile infiltration, spinal cord injury, spinal anaesthesia or general anaesthesia.
2. **High flow:** Perineal or penile trauma.

Two types

Type I: Low flow priapism (veno-occlusive)
Decreased venous outflow
Increased intracavernosal pressure
Painful, fully erect penis
Local hypoxia and acidosis
It is the most common variety

Type II: High flow priapism
High inflow and high outflow
Penis is erect but nontender
Corporal blood gas analysis can differentiate low flow and high flow priapsim.

Treatment

1. **Low flow priapism:** Corporal irrigation with normal saline and α-adrenergic intracorporal injections (epinephrine/phenylephrine/ephedrine) every 5 min till detumescence.
   In severe cases—shunts (corporoglandular), corporospongiosal or corporosaphenous) may be necessary. If patient has sickle cell disease, IV bicarbonate and blood transfusions required.
2. **High flow priapism:** Doppler ultrasound done to identify arterial-lacunar fistula.
   Arterial embolisation or open surgical arterial ligation done.

INTERESTING 'MOST COMMON' FOR TESTICULAR CANCER

- Most testicular tumours are malignant
- Most common cancer in young adults is testicular cancer
- Most common solid cancer in young man is seminoma
- Most common tumour is seminoma
- Most common teratoma is malignant teratoma intermediate: Teratocarcinoma

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All the topics have been updated
- New photographs and key boxes have been added

MULTIPLE CHOICE QUESTIONS

1. Following are true for surgical anatomy of the penis *except*:
A. Corpora cavernosa are vascular spaces
B. Arterioles are corkscrew shaped
C. Attached to symphysis pubis by suspensory ligament
D. Deep artery supplies corpus spongiosum alone

2. Balanitis refer to:
A. Inflammation of the glans penis
B. Inflammation of the prepuce
C. Inflammation of glands in the fossa navicularis
D. Inflammation of the urethral glands

3. Dorsal slit is given for which condition?
A. Phimosis
B. Paraphimosis
C. Carcinoma penis
D. Balanitis

4. Following are complications of phimosis *except*:
A. Paraphimosis
B. Carcinoma penis
C. Balanoposthitis
D. Buschke-Lowenstein tumour

5. Which of the following is rare feature of carcinoma penis?
A. Foul smelling discharge
B. Recent phimosis
C. Urethral involvement
D. Induration is extensive

6. Treatment of carcinoma penis confined to prepuce is:
A. Circumcision
B. Dorsal slit and excision
C. Partial amputation
D. Radiotherapy

7. Following are true for partial amputation of penis *except*:
A. Partial amputation is the treatment for growth confined to glans penis
B. At least 2 cm proximal shaft is necessary in partial amputation
C. Long ventral flap is required to cover
D. Perineal urethrostomy may be required

8. Few important consideration sin total amputation of the penis include following *except*:
A. Suspensory ligaments have to be divided
B. Perineal urethrostomy has to be done
C. Severe excoriation of the scrotum is a complication
D. Advantage being no stricture urethra after surgery

9. Following are the features syphilitic chancre of the penis *except*:
A. Single ulcer
B. Hard chancre
C. Firm painful lymph nodes in the axilla
D. Contagious

10. Testosterone is produced by?
A. Germ cells B. Sertoli cells
C. Leydig cells D. Clear cells

11. Testicular artery is a branch of:
A. Aorta
B. Common iliac
C. Internal iliac
D. Early morning urine sample is the best to measure this

12. Following are true for prostate specific antigen *except*:
A. Released from columnar prostatic acinar epithelial cells
B. More than 4 nmol/ml suggest carcinoma prostate
C. Prostatitis also can increase in the levels
D. It does not help in assessing the response to treatment

13. Which of the following test differentiate encyste hydrocoele from spermatocoele?
A. Fluctuation
B. Getting above the swelling
C. Impulse on cough
D. Traction test

14. What is the classical clinical feature of tuberculou epididymo-orchitis?
A. Thickened vas
B. Atrophy of the testis
C. Craggy epididymis
D. Anterior sinus

15. Chinese lantern type of transillumination is classica of:
A. Spermatocoele B. Epididymal cyst
C. Ranula D. Cystic hygroma

16. Which of the following is true in cases of retractil testis?
A. It is premalignant
B. Testis is generally not palpable
C. Scrotum is well developed
D. Testis cannot be brought down into scrotum

17. In torsion testis following are true *except*:
A. Scrotum is empty
B. Tender lump at external abdominal ring
C. Elevation of scrotum increases pain
D. Upper abdominal pain and vomiting are features

18. Which one of the following is more prone for testicular tumour?
A. Gardner's syndrome
B. Down's syndrome
C. Klinefelter's syndrome
D. Sipple syndrome

19. Human chorionic gonadotrophin elevation is diagnostic of .....
A. Choriocarcinoma
B. Seminoma
C. Leydig cell tumour
D. Sertoli cell tumour

20. Following are true for increased LDH levels in tumours:
A. It suggests liver metastasis
B. It suggests germ cell tumours
C. It suggests malignant melanoma
D. It suggests tumour load

ANSWERS

1 D	2 A	3 B	4 D	5 C	6 A	7 D	8 D	9 C	10 C
11 A	12 D	13 D	14 C	15 B	16 C	17 D	18 C	19 A	20 B

43

Differential Diagnosis of Haematuria

- Causes
- History and examination
- Investigations
- Haematuria
- History
- What is new?/Recent advances

CAUSES OF HAEMATURIA (Fig. 43.1 and Key Box 43.1)

I. In the kidney

1. Infection
- Acute glomerulonephritis
- Tuberculosis

2. Infarction
- SBE with emboli causing renal infarction
- Massive haemolysis with acute renal tubular necrosis
- Mismatched blood transfusion

3. Injury
- Stab/blunt injury

4. Tumours
- Wilms' tumour: Nephroblastoma
- Hypernephroma: Renal cell carcinoma (RCC)
- Transitional cell carcinoma (TCC)

5. Stones

6. Polycystic kidney

KEY BOX 43.1

HAEMATURIA

Common causes
- Urolithiasis
- Tumours

Uncommon causes
- Tuberculosis
- Cystitis, bladder tumours
- Polycystic kidney

II. In the ureter

1. Stone
2. Cancer—rare

III. In the urinary bladder

1. Carcinoma of bladder
2. Carcinoma prostate
3. Cystitis
4. Tuberculosis
5. Bilharziasis
6. Stone: Common in school-going children
7. Benign prostatic hyperplasia (BPH)

IV. Urethra

1. Stone

V. Rare causes

1. Patients on anticoagulants
2. Sickle cell anaemia
3. Bleeding disorders

HISTORY AND EXAMINATION

1. Age and sex

Young children	Vesical calculus
Young adults	Renal stones, tuberculosis (TB)
Elderly patients	Renal cell carcinoma (RCC)

2. Occupation

Aniline dye workers	Carcinoma bladder

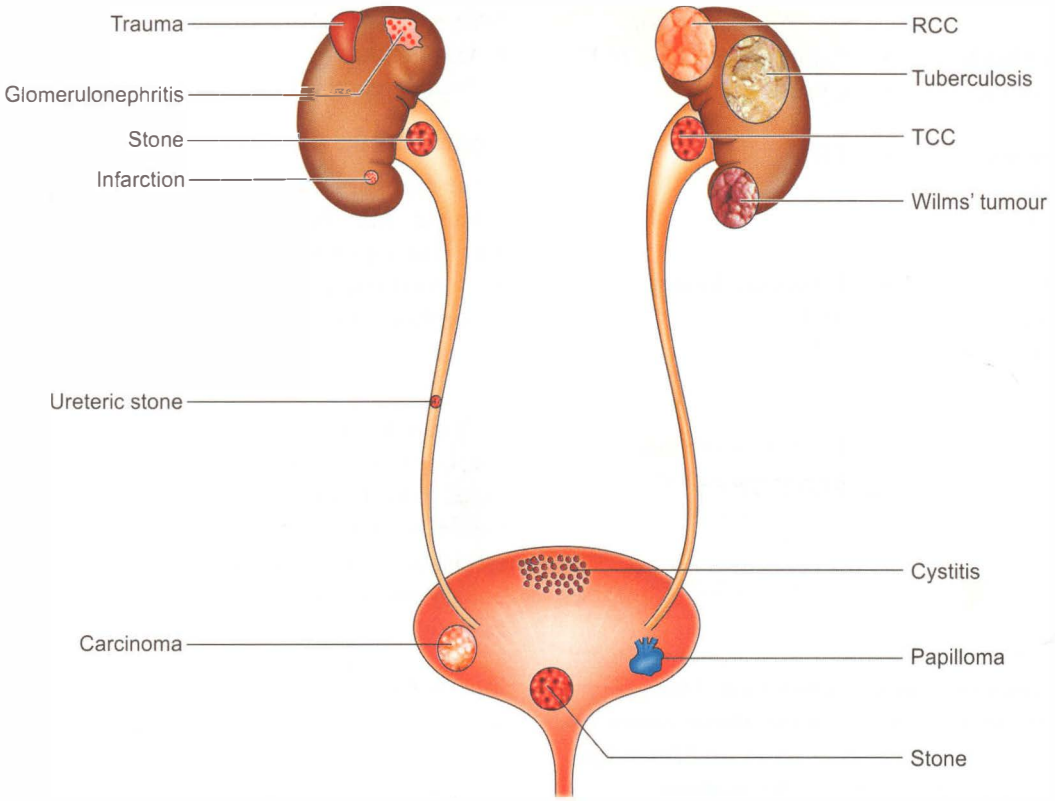


Fig. 43.1: Various causes of haematuria

3. Haematuria

• Bright red	• Lower urinary tract
• Altered blood	• Kidney
• Profuse	• Papilloma
• Small quantity	• Renal cell carcinoma, TB, stone
• Beginning of micturition	• Urethral pathology
• End of micturition	• Bladder pathology
• Mixed with urine	• Renal
• Painless haematuria	• Papilloma or carcinoma
• Painful haematuria	• Renal stone, bladder stone

4. General physical examination

• Gross pallor	• Significant blood loss
• Gross pallor with minimal blood loss	• Renal cell carcinoma
• Hypertension	• Polycystic kidney
• Bony pains	• Carcinoma (prostate)

5. Abdominal examination

• Palpable kidney	• Polycystic kidney, Wilms' tumour, renal cell carcinoma
• Distended bladder	• Carcinoma prostate, enlarged prostate
• Suprapubic tenderness	• Bladder stone, cystitis
• Craggy epididymis and beaded vas	• Genitourinary tuberculosis

6. Rectal examination

• Enlarged smooth prostate	• BPH
• Hard irregular prostate	• Carcinoma prostate
• Hard, thickened seminal vesicles	• Genitourinary tuberculosis
• Advanced growth	• Carcinoma rectum infiltrating urinary bladder

INVESTIGATIONS (Table 43.1 and Fig. 43.2)

1. Urine examination

• Worm-like clots	• Growth in the ureter
• Flat disc-like	• Urethra
• Pieces of tumour	• **Papilloma** of the bladder

2. Urine microscopy

• Pus cells	• Urinary tract infection
• Abacterial acid pyuria	• TB
• Malignant cells positive	• RCC or **papilloma** bladder

3. Plain X-ray KUB

• Enlarged kidney	• Polycystic kidney, renal cell carcinoma
• Radio-opaque shadows	• Renal stones, ureteric stones, bladder stone

4. Cystoscopy

- Growth in the bladder
- Inflammation of the bladder
- Ulcers, hyperaemia, golf-hole ureter
- Papilloma bladder/TCC
- Cystitis
- TB

5. Intravenous urography

- Spider leg calyces
- Irregular calyces
- Missing calyces
- Polycystic kidney
- RCC
- TB

6. Ultrasound

- Enlarged kidney
- Renal cell carcinoma
- Polycystic kidney
- Wilms' tumour
- Stones

PEARLS OF WISDOM

Differential diagnosis of haematuria is a major theory question in an undergraduate examination. I have given some ideas about how to evaluate these cases by analysing history, physical examination and investigations. This gives a good exercise to the students.

HAEMATURIA

Haematuria is defined as passage of blood mixed with urine.

It can be classified as

1. Microscopic
2. Macroscopic

Also
1. Glomerular
2. Nonglomerular (urological—Table 43.2).

Macroscopic or gross haematuria

Urine may vary in colour from red to stale brown or chocolate coloured. May be associated with clots. The nature of clots may give a clue to the source or site of bleeding. Serpiginous or vermiform clots indicate bleeding from the kidneys. Amorphous clots are suggestive of lower tract bleeding. Most patients with gross haematuria have some pathology in the kidneys or the genitourinary tract.

It can be initial, terminal or throughout the stream. Urethral bleeding usually manifests as the first 10–15 ml of blood-stained urine, which gradually clears. Terminal haematuria usually indicates trigonal irritation (e.g. bladder stone). It is usually associated with dysuria and strangury. Bleeding from the upper urinary tract (kidneys and ureters) is seen throughout the stream as the blood mixes with the urine stored in the bladder.

Microscopic haematuria

It is defined as the presence of more than 3 RBCs per high power field on microscopic examination. It is usually detected incidentally on urine dipstick testing and subsequent microscopic examination.

Normal RBC excretion is up to 2 million RBCs/day.

Aetiology

Evaluation: Numerous investigative modalities are available for evaluation of haematuria. The choice of the correct modality depends on the clinical suspicion and important clues on clinical examination.

Table 43.1 Utility of various investigative modalities for evaluation of haematuria

Investigations	Advantages	Disadvantages
Intravenous urography (IVU)	Evaluation of kidney collecting system and ureter for stones, masses and obstruction	Poor assessment of bladder pathology Nephrotoxicity
Cystoscopy	Investigation of choice for bladder and urethral assessment	Invasive Significant patient discomfort Expensive equipment required Potential for complications (e.g. urethral stricture)
Urinary cytology	Diagnosis of transitional cell carcinoma (TCC) Grading of TCC	Not useful for squamous or adenocarcinoma
CT scan	Investigation of choice for colic (noncontrast CT scan) Best investigation for renal masses Better than IVU in most situations Other organs affecting the urinary tract can also be imaged (e.g. carcinoma cervix causing ureteric obstruction)	Cost Radiation Contrast nephrotoxicity
Angiography	Aneurysms A-V fistulas/malformations Postoperative bleeding for localisation and angioembolisation	Invasive Not widely available (skilled) High cost

Initial evaluation includes

Ultrasonography: It is noninvasive and less expensive

- Diagnosis of upper or lower urinary tract lesions
  a. Stones b. Tumours c. Hydronephrosis
- Renal parenchymal changes suggest glomerular cause.
- Ultrasound guides towards choosing further investigations
- Less sensitive for evaluation of ureteral causes of haematuria.

Table 43. 2 Distinguishing between glomerular and urological causes of haematuria

	Glomerular	Nonglomerular
Colour (if macroscopic)	Pale red, smoky brown or stale tea	Red or pink
Clots	Rare	More common
Proteinuria (dipstick/mg per day)	3+ or more (> 500 mg/day)	< 2+ (< 500 mg/day)
RBC morphology	Dysmorphic RBCs	Normal
RBC casts	May be present	Absent

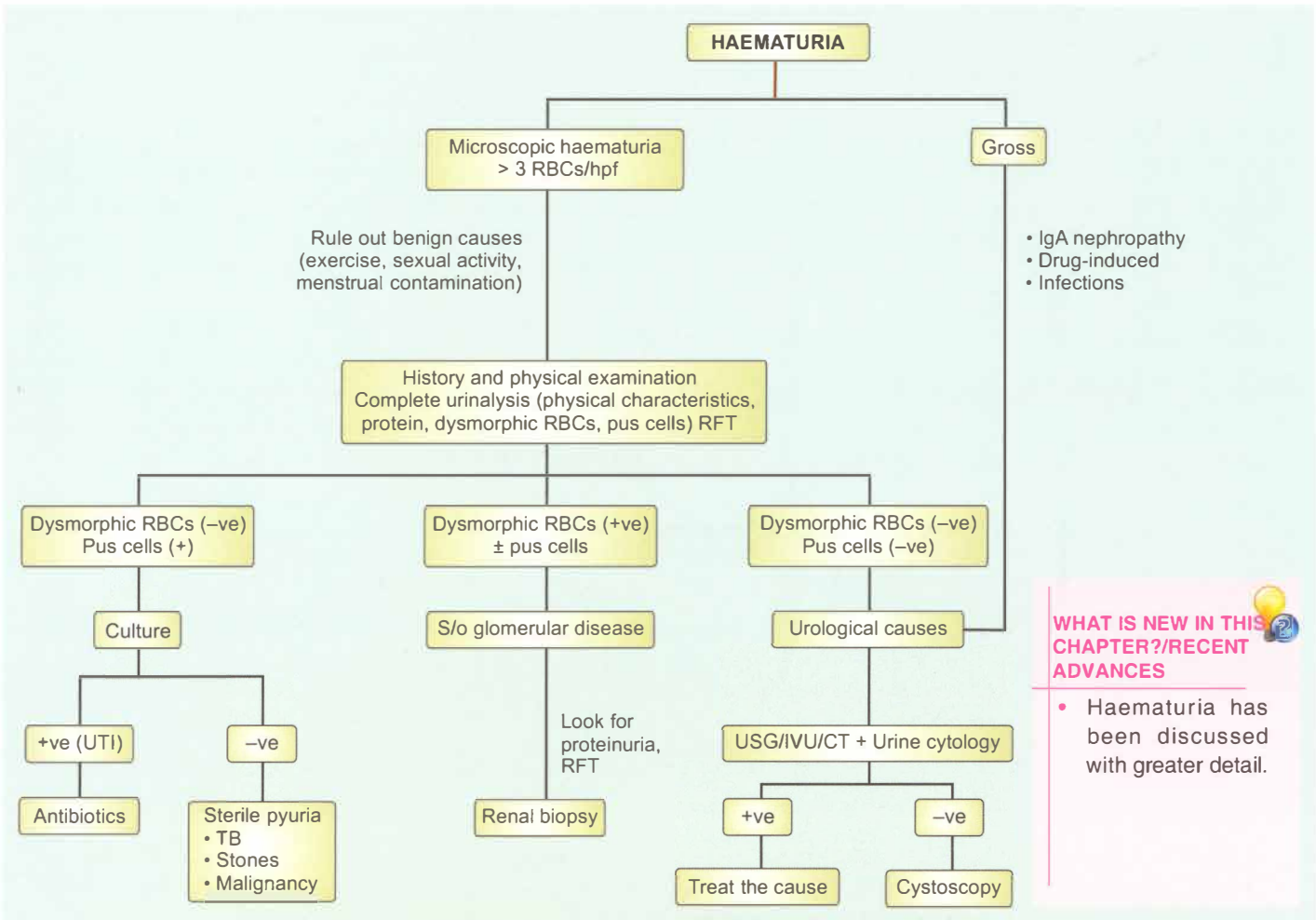


Fig. 43.2: Haematuria—management algorithm

MULTIPLE CHOICE QUESTIONS

1. Following is the cause of haematuria in bacterial endocarditis:

A. Renal necrosis B. Renal sepsis
C. Renal infarction D. Coagulopathy

2. Haematuria in polycystic kidney is due to:

A. Hypertension B. Renal infarction
C. Renal ischaemia D. Cyst rupture into renal pelvis

3. Which of the following conditions rarely gives rise to haematuria?

A. Renal stones
B. Renal cell carcinoma
C. Bilharziasis
D. Benign prostatic hypertrophy

4. Palpable renal masses in 40-year-old hypertensive patient with one attack of haematuria is:

A. Hypernephroma
B. Hydronephrosis
C. Adenomyolipoma
D. Polycystic kidney

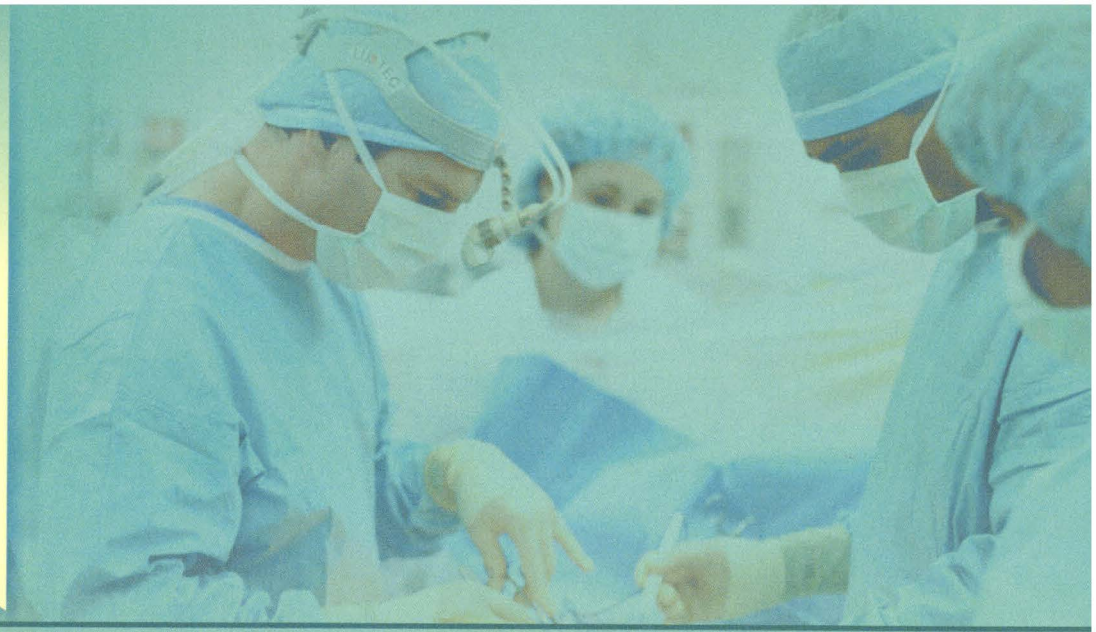
5. Which of the following pathologies is reflected as terminal haematuria?

A. Trigonal irritation
B. Posterior urethral irritation
C. Ureteric irritation
D. Renal irritation

ANSWERS

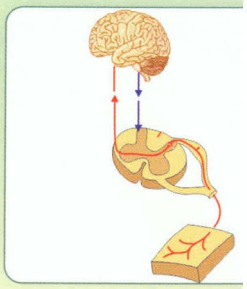
1 A 2 D 3 D 4 D 5 A

Section
IV



Specialities

44. Chest Trauma, Cardiothoracic Surgery
45. Neurosurgery
46. Principles of Radiology
47. Principles of Clinical Radiation Oncology
48. Principles of Anaesthesiology
49. Organ Transplantation



44

Chest Trauma, Cardiothoracic Surgery

- Chest trauma
- Blunt trauma
- Pulmonary injuries
- Tracheobronchial injuries
- Myocardial contusion
- Surgical emphysema
- Mediastinal emphysema
- Mediastinal masses
- Pulmonary aspergilloma
- Congenital heart diseases
- Patent ductus arteriosus
- Coarctation of aorta
- Coronary artery bypass graft
- Off pump coronary artery bypass surgery
- Abdominal aortic aneurysms (AAA)
- What is new?/Recent advances

CHEST TRAUMA

Introduction

In chest trauma, the mortality is very high unless promptly recognised and properly treated. The margin of safety is very slim, initial care dictates the final result. With varying degree of severity, chest injuries occur in almost 80% of road traffic accidents (Key Box 44.1).

MAIN AIMS OF RESUSCITATION

The standard method of resuscitation in all cases of polytrauma is as follows:

A. Airway

- Aspiration of blood and secretions from oral cavity, pharynx and trachea
- Introduce plastic airway
- Endotracheal intubation

KEY BOX 44.1

COMMON CAUSES

- Automobile accidents
- Gunshot wounds
- Stab injuries
- Blast injuries
- Crush injuries

- Cricothyroidotomy as necessary
- Tracheostomy as necessary

B. Breathing

- Intubation
- Intercostal chest tube (ICT)
- Closure of any open chest wounds

These two steps (A and B) help in re-aerating the lung

C. Circulation

- Control of major and life-threatening bleeding
- Volume infusion

D. Disability

- Neurological

E. Exposure

- All clothing to be cut open without moving the patient
- All the above steps are taken by the trauma centre team simultaneously and not one by one.
- Relieve pain. Do not sedate
- All open wounds of the chest to be covered
- Life-threatening injuries should be identified and treated immediately.

ASSESSMENT OF INJURY

History

- Time since the injury
- Details of the injury from the bystander or the police

- High-speed deceleration injury (aortic and cardiac rupture to be ruled out).
- Crushing accidents (tracheobronchial and oesophageal tear).
- Sudden abdominal compression (ruptured diaphragm).
- In stab injuries, length of the knife and direction of stab.

Examination

- The clothing should be removed carefully without moving the patient.
- Palpate for clinical evidence of fracture ribs, surgical emphysema, any paradoxical movement of the ribs and auscultate for air entry in both lungs.
- Even if it is a trivial injury, the patient should be admitted and observed for a minimum of 24 hours before discharge.
- It is reasonable to do unilateral or bilateral closed tube thoracostomy (ICT) (Fig. 44.1) on suspicion of haemothorax or pneumothorax when the patient is in respiratory distress.

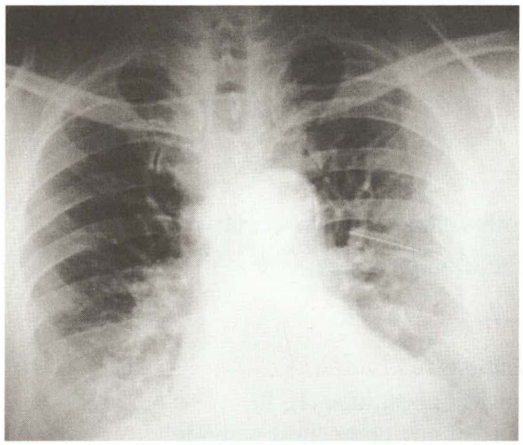


Fig. 44.1: Fracture ribs with haemothorax left side—intercostal tube has been inserted

BLUNT TRAUMA TO THE CHEST

Causes

- Road traffic accidents
- Fall from a height
- Crush injuries
- Assault with blunt object

SIMPLE RIB FRACTURE

Rib fracture can be single or multiple (Key Box 44.2).

Single rib fracture

- Often regarded as a trivial injury but should be treated with respect in elderly patients.
- Occurs due to direct injury or excessive flexion.
- The common site is at the costal angle or middle of the shaft.
- Patients will have pain on breathing, coughing and on palpation.

KEY BOX 44.2

POINTS TO REMEMBER

- First rib fracture is a marker of severe trauma—injury to brachial plexus, subclavian artery and vein
- Displaced fracture of 8th–10th ribs — Injury to liver and spleen
- Fracture of 11th and 12th ribs—injury to kidney
- Penetrating injury to left lower chest wall—injury to heart, lung, diaphragm, stomach and spleen

- They are treated with analgesics, intercostal nerve block and assurance.

Multiple rib fractures

- When there are multiple rib fractures without any pneumo- or **haemothorax** and no other organs are involved, intercostal nerve block and small amount of narcotics are required.
- Strapping is occasionally necessary in young adults.
- In elderly patients, consider hospitalisation for observation, pain control and pulmonary toilet.
- Chest X-ray to be repeated after 24 hour and at the time of discharge, to rule out late onset pneumo- and haemothorax.
- Intermittent use of velcro belt rib support.
- Inform the patient of deep breathing and coughing using the rib belt.
- Epidural analgesia is becoming the standard of care for pain management in patients with multiple rib fractures.

STERNAL FRACTURE

- Commonly due to steering wheel injury—blunt trauma.
- Usually occurs at the sternal angle.
- Associated with costochondral dislocations.
- Classified as **displaced and nondisplaced fractures**.
- Localised swelling, tenderness and deformity are the clinical findings.

Treatment

- Displaced fracture—requires surgical fixation.
- Nondisplaced fracture—conservative management.

FLAIL CHEST

This results from severe chest injuries with multiple rib fractures.

Here there are fracture of **four or more ribs at two places,** anteriorly and posteriorly, so that certain segments of ribs will have no attachment to the chest wall. These ribs become indrawn due to intrathoracic negative pressure as the patient inhales and is driven outwards on expiration producing instability. This is called **paradoxical respiration.** It results in hypoventilation, carbon dioxide retention and respiratory failure.

Flail chest is of three types: Anterior, posterior and lateral.

Anterior flail

- Fracture of the costochondral junction on both side of the sternum

Posterior flail

- Fracture ribs of posterior chest wall

Lateral flail

- Fracture shaft of the ribs

Treatment

I. Commonly done procedures

1. **Anterior flail:** Seagull-shaped prosthesis introduced to stabilise the flail segments.
2. **Posterior flail:** No treatment is required as the scapula acts as a support to stabilise the flail segment.
3. **Lateral flail:** It is treated by chest wall stabilisation, reduction of the respiratory dead space, management of the pulmonary contusion and pain control. Epidural analgesia is recommended for pain management. Intercostal nerve blocks may also be used.

II. Other methods

- Surgical stabilisation is rarely indicated, i.e. open reduction of rib fracture or osteofixation.
- Recent method is to intubate and stabilise the flail segments with positive pressure ventilation, which has to be done for at least one week. This is called internal pneumatic fixation.
- Physiologic stabilisation with intubation and IPPV must be initiated before hypoxia develops. IPPV produces satisfactory ventilation and helps the fractured ribs to unite in the position of inspiration, thereby reducing the deformity and improving the pulmonary function.

'STOVE IN' CHEST

- Localised, severe, blunt or crush injury produces depression of a portion of the chest.
- Treatment is same as that for flail chest. Sometimes, thoracotomy may be needed if there are internal injuries.

PULMONARY INJURIES

CONTUSION OF THE LUNG

- Deceleration injuries or crush trauma often produces extensive parenchymal damage. Haemorrhage and interstitial oedema result in obliteration of alveolar spaces and consolidation of large areas of pulmonary tissue.
- Contusion of the lung can be unilateral or bilateral. The contusion can be in the form of a small area of damage with oedema and extravasation of the blood, or it may be widespread damage. Haemoptysis and excessive tracheobronchial secretions give the clue to the diagnosis (Key Box 44.3).

KEY BOX 44.3

PULMONARY INJURIES

- Contusion
- Pneumothorax/haemothorax
- Laceration
- Chest wall injuries

- Chest X-ray: Early patchy consolidation. It must be differentiated from adult respiratory distress syndrome (ARDS).
- CT scan is more specific (Fig. 44.2).

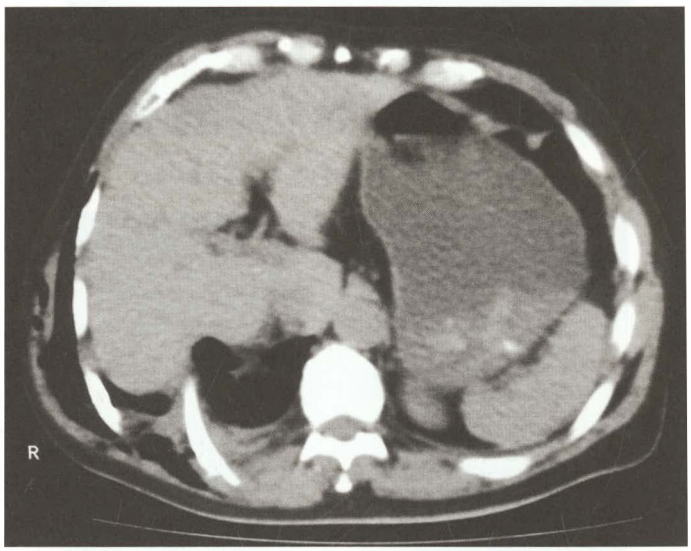


Fig. 44.2: CT scan showing rib fracture and lung injury

Treatment

- Fluid restriction, pulmonary care
- Chest physiotherapy
- Steroids and rarely ventilation

Usually self-limiting, if there are no other severe injuries.

Complications

- Pneumonia
- Atelectasis
- Respiratory failure
- ARDS

PNEUMOTHORAX

- Pneumothorax is the most common cause of respiratory insufficiency following chest trauma.
- Usually if there is a rib fracture and evidence of subcutaneous emphysema, pneumothorax is certainly present (Fig. 44.3).
- Pneumothorax can be closed (simple), open and tension.
- Small simple pneumothorax does not need any treatment.
- A repeat chest X-ray after 12–24 hours is essential to confirm that it is not progressing.

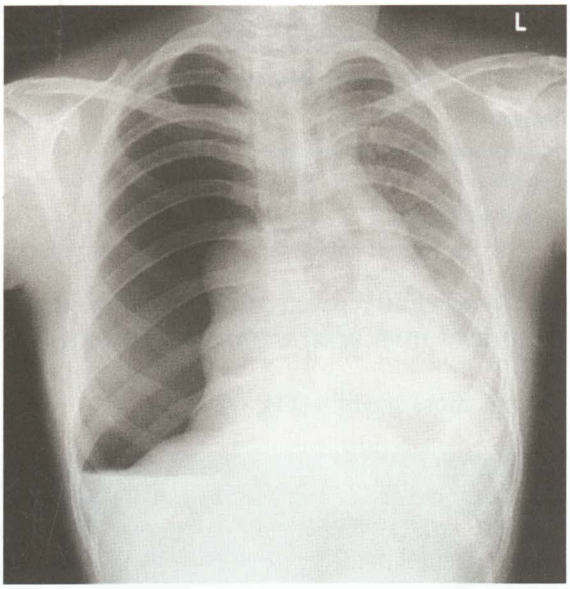


Fig. 44.3: Pneumothorax right lung

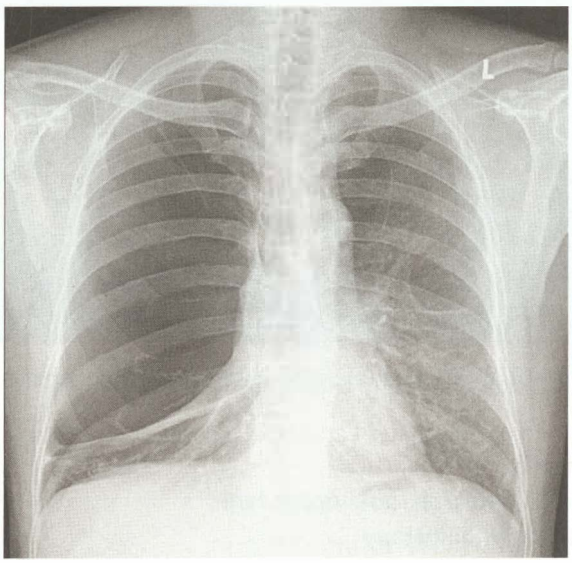


Fig. 44.4: Large bullae

- It can be confused for a large bullae (Fig. 44.4).
- Small pneumothorax can be missed easily.
- **Bilateral pneumothorax is an emergency**.
- Late pneumothorax can also occur.
- Open chest wound will produce complete collapse of lung and paradoxical shift of the mediastinum with each respiration (mediastinal flutter) causing hypoventilation and reduced cardiac output.
- Treatment is by closure of the wound, intercostal tube drainage (ICD) and surgery.

Tension pneumothorax (Key Boxes 44.4 to 44.6)

- Injury to the lung results in continuous valvular air leak. The accumulating air collapses the lung on the same side and pushes the mediastinum to opposite side. As a result of this tension, the intrapleural pressure increases, till it is above atmospheric pressure at the time of expiration. This **reduces the venous return** to the heart as well as compromises the ventilation.
- Tension pneumothorax is **an emergency** which should be treated urgently with needle thoracocentesis in the second intercostal space in the midclavicular line to release the tension. **Thoracocentesis** converts tension pneumothorax to simple pneumothorax.
- This should be followed by urgent ICD insertion and connected to underwater seal.
- **Do not wait for chest X-ray**.

KEY BOX 44.4

TENSION PNEUMOTHORAX: CLINICAL FEATURES

Tachypnoea
Tachycardia
Tympanic note on percussion
Total absence of breath sounds
Tracheal shift *Observe* **5 Ts**

KEY BOX 44.5

TENSION PNEUMOTHORAX

Injury to the lung
↓
Valvular air leak
↓
Lung collapse
↓
Mediastinal shift to opposite side
↓
Increased intrapleural pressure
↓
Decreased venous return
↓
Decreased ventilation
↓
Hypoxia and cardiac arrest

KEY BOX 44.6

TENSION PNEUMOTHORAX

- Diagnosis immediately
- Do not wait for CXR
- Thoracocentesis in 2nd intercostal space, midclavicular line.
- Then, follow with intercostal chest tube insertion.

Tension gastrothorax

This occurs due to herniation of dilated and obstructed stomach into the mediastinum due to diaphragmatic tear resulting in haemodynamic compromise. It should be treated by reducing the hernial contents and repair of diaphragmatic tear.

HAEMOTHORAX

- May be missed in chest X-rays in the supine position.
- Results from injury to internal mammary artery, intercostal artery and vascular lung adhesions.
- **Classical signs are** reduced—chest expansion, dullness to percussion and absent breath sounds on affected side.
- Treated by intercostal chest tube insertion (Figs 44.5 and 44.6).
- If bleeding continues or features of shock develop, thoracotomy has to be considered.
- The bleeding may be delayed or may recur after several days.

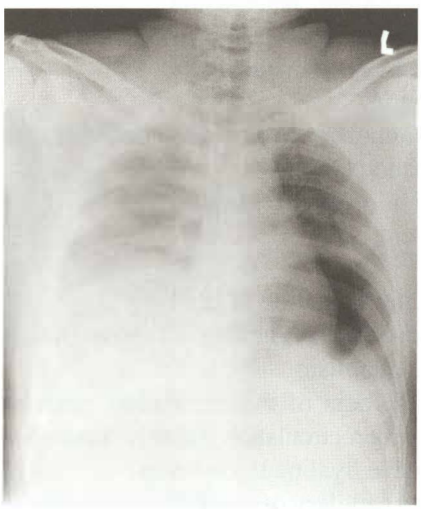


Fig. 44.5: Right haemothorax

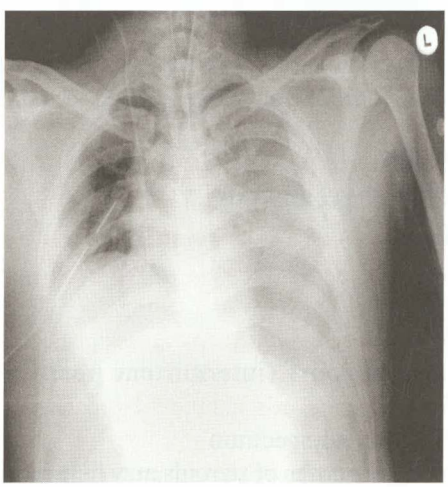


Fig. 44.6: Bilateral haemothorax—detected early and treated with ICT on the right

Indications for thoracotomy

- Initial volume of blood loss is not as important as the amount of ongoing bleeding.
- Drainage is more than 1000 ml or 100 ml each hour for 4 hours.
- If clotted haemothorax is suspected (opacity persisting on chest X-ray even after ICD).

Complications

Failure to adequately drain a haemothorax initially results in residual clotted haemothorax and empyema or late fibrothorax.

INTERCOSTAL CHEST TUBE (ICT) INSERTION (CLOSED TUBE THORACOSTOMY) (Flowchart 44.1)

- **Second intercostal space**—anteriorly midclavicular line is ideal for pneumothorax and sixth space in the midaxillary line for haemothorax.
- **Triangle of safety**
  - Above the level of nipples
  - Anterior to midaxillary line
  - Below and lateral to pectoralis major muscle
- Can be introduced from sixth intercostal space midaxillary line for pneumothorax also but the chest tube should reach the apex of the lung (Figs 44.7 and 44.8).
- **Infiltrate local anaesthetics** up to the parietal pleura
- 2–3 cm incision parallel to the ribs—deepened, suture taken
- Insert the chest tube with trocar into the pleural cavity
- Then the trocar is removed and the chest tube is connected to underwater seal. The ICT is fixed
- Intercostal drainage can also be done by connecting to 2 bottles (Fig. 44.9)
- Removal of ICT (Key Box 44.7)

For ten commandments of ICT—*see* the next page.

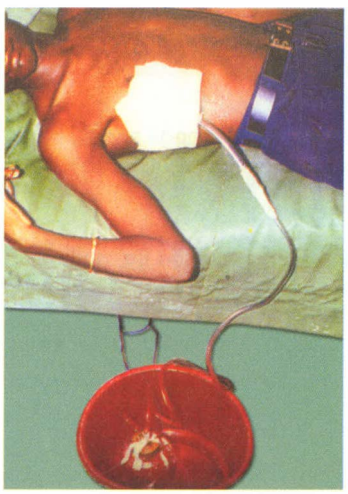


Fig. 44.7: Intercostal tube (ICT) connected to underwater seal

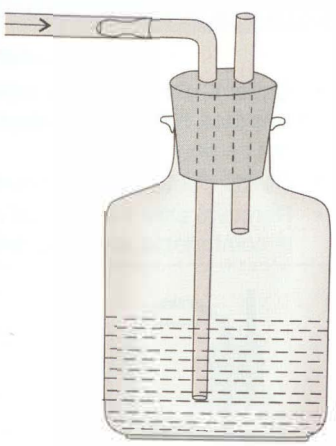
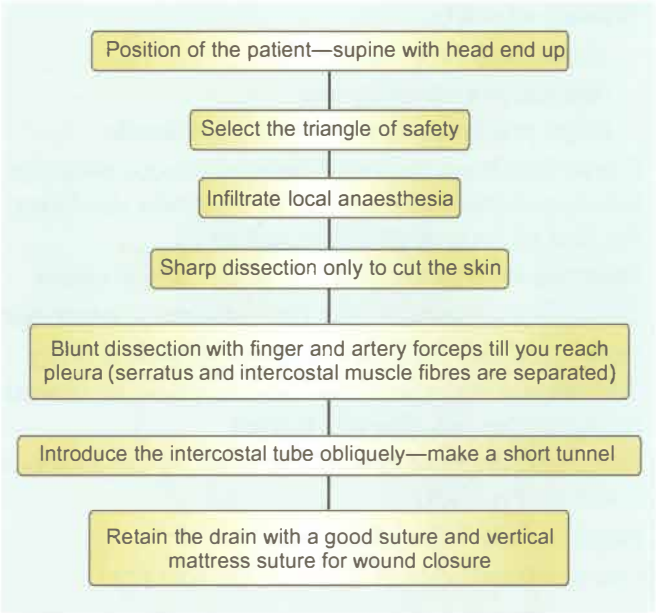


Fig. 44.8: Underwater seal (diagrammatic)

TEN COMMANDMENTS OF PRECAUTIONS WHILE USING ICT–ICD

1. Should select triangle of safety
2. Should direct the tube towards apex in cases of haemothorax and pneumothorax
3. Should direct the tube towards base in empyema
4. Should confirm that all the holes are inside
5. Should be connected to under water seal
6. Should observe for movement of the fluid column with respiration
7. Should take an X-ray after the insertion of the tube
8. Should avoid kinking of the tube
9. Should not clamp the ICD if there is air leak
10. Should be always lower than patient level in the supine position. Otherwise, contents of the bottle will enter the pleural space

Flowchart 44.1: Important steps of intercostal tube insertion



KEY BOX 44.7

ICT—REMOVAL

- Chest X-ray—lung fully expanded and drainage should be less than 100 ml for two days and no air leak. Patient should not be on a ventilator
- Clamp the tube for 24 hours
- Removal after 24 hours of clamping, provided the patient is comfortable and lung remains fully expanded.

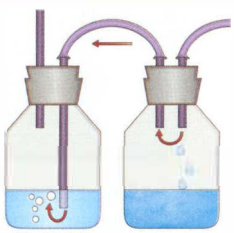


Fig. 44.9: ICD with 2 bottles

LUNG LACERATION

- **Minor laceration:** Haemopneumothorax. Usually inter costal chest tube is enough.
- **Major laceration:** Haemopneumothorax
  i. Introduce intercostal chest tube
  ii. Continuous air leak or bleeding through ICT and lung not expanding. Requires thoracotomy and repair or resection of lobe.

SHOCK LUNG

Definition

- Alveolar collapse due to shock as a result of oedema, impaired perfusion, and reduction in alveolar space resulting in respiratory failure is called shock lung.
- It is also called **A**cute **R**espiratory **D**istress **S**yndrome (**ARDS**).

Causes

- Major chest trauma with multiple rib fractures and lung contusion
- Septic shock and septicaemia
- Disseminated intravascular coagulation
- Massive blood transfusion
- Major burns
- Cardiopulmonary bypass (also called 'Pump lung')
- Acute pancreatitis
- Aspiration of gastric contents

Pathogenesis

- Diffuse inflammation of the lung
- Extensive intravascular coagulation due to micro-thromboembolism
- Focal disorders of the circulation, primarily because of sluggish microcirculation due to leukostasis, sludge, leading to extensive hyaline thromboses
- Increased capillary permeability due to damaged capillaries
- Diffuse alveolar damage, decreased production of surfactant by type II pneumocytes leading to bilateral extensive fine atelectasis
- Net result is pulmonary consolidation, decrease in the lung compliance, poor gas exchange leading to stiff lung.

Investigations

- **Arterial blood gas analysis**
- **Chest X-ray**
- **CT scan**

Treatment

- Ventilatory support (intermittent positive pressure ventilation)
- Antibiotics—broad spectrum
- Low to moderate doses of steroids may help in early ARDS, in patients requiring high doses of vasopressors to maintain blood pressure.

Table 44.1 Summary of blunt chest injury

Rib fractures	Pulmonary injuries	Tracheobronchial	Diaphragm
Flail chest	Contusion	Mediastinal emphysema	Breathlessness
Stove-in chest	Pneumothorax	Shock	Coils of bowel within thorax
Subclavian injury	Haemothorax	Airway obstruction	
Surgical emphysema	Laceration	Death	

- Intensive care—supportive therapy can be remembered as **FASTHUG**
  - **F**—Feeding (usually enteral)
  - **A**—Analgesia
  - **S**—Sedation once a day and check neurological status
  - **T**—Thromboembolism prophylaxis
  - **H**—Head-end elevation (20–30°)
  - **U**—Ulcer (gastric) prophylaxis, and
  - **G**—Glucose (blood) control

INJURY TO TRACHEA AND MAJOR BRONCHI

TRACHEAL INJURIES

Injuries occur as a result of trauma sustained due to crush injuries. Common in cervical trachea.

Symptoms

- Acute airway obstruction
- Emphysema—mediastinal and cervical
- Pneumothorax voice impairment

Treatment

Intubation, tracheostomy, surgery and repair

MAJOR BRONCHIAL INJURIES

- Blunt trauma produces stereotype injury to main bronchus of either side. Lobar bronchi are less commonly injured (Table 44.1).
- Lesion is often a circumferential laceration with complete separation or a partial tear.

Symptoms

- Pneumothorax, uncontrolled air leak
- Haemoptysis
- Surgical emphysema
- Bronchoscopy for confirmation

Surgery

- Endobronchial intubation
- Bronchoplasty, injury to the lobar bronchus—resection of the lobe of the lung.

INJURY TO THE DIAPHRAGM

Features

- Due to crush injury
- Industrial—due to fall of heavy weights over the abdomen.
- Commonly, left diaphragm tear occurs.
- Missed in chest X-ray in the supine position.
- Diagnosed by CECT of the abdomen or chest X-ray reveals the Ryle's tube in the chest or coils of bowel (gas shadow) in the chest and elevated fundic air bubble (Figs 44.10 to 44.12). Diagnostic laparoscopy is also a good investigation in cases of rupture diaphragm.
- When in doubt, do diagnostic laparotomy.

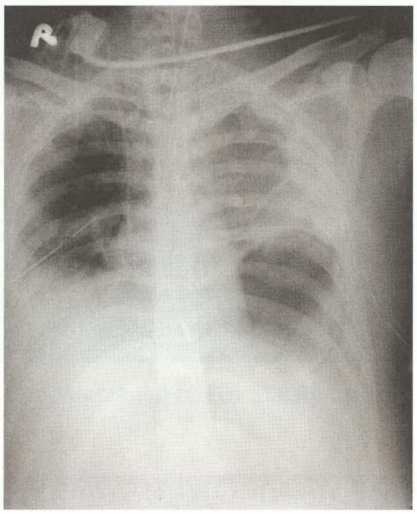


Fig. 44.10: Chest X-ray left haemothorax with elevated fundic air bubble

INJURY TO THE AORTA (RUPTURE OF THE AORTA)

- Deceleration injuries
- Gets avulsed at the region of ligamentum arteriosus
- Complete rupture—immediate death
- Incomplete rupture—shock, widening of mediastinum, unequal pulses.

Surgery

- Repair is done using cardiopulmonary bypass.

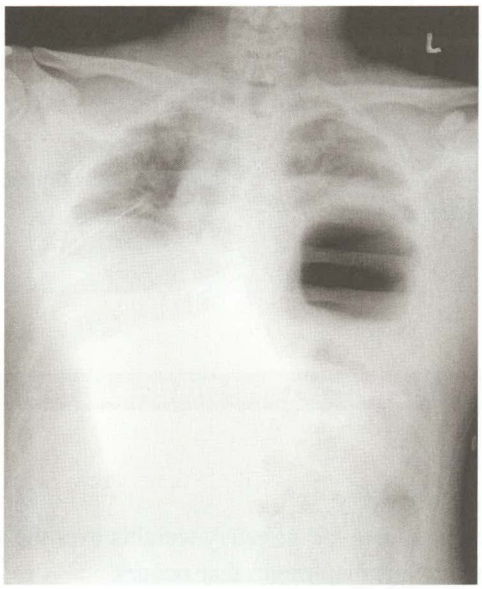


Fig. 44.11: Fundic air bubble in the thorax

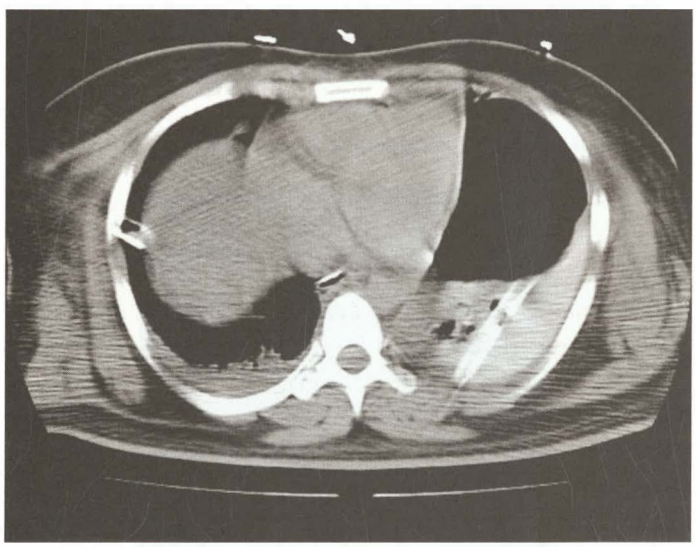


Fig. 44.12: ICT in the stomach

MYOCARDIAL CONTUSION

- Deceleration injuries
- Anterior chest impact
- Clinical feature include arrhythmias, reduced cardiac output, cardiac tamponade.

SURGICAL EMPHYSEMA

Types

1. Localised
2. Extensive from the eyelids to the scrotum, quite alarming in appearance. Indicates lung injury (Fig. 44.13).

PEARLS OF WISDOM

Palpable crepitus is diagnostic of surgical emphysema.

X-ray chest may show pneumothorax or air in the subcutaneous plane (Fig. 44.14).

Treatment

- **Localised:** Not extending, no pneumothorax—observation
- Pneumothorax—ICT
- **Extensive:** ICT to be introduced on the side where it is maximum.
- ICT may be required on both sides
- Two ICT, one apical and the other, basal, may need to be introduced if there is continuous air leak.

Fig. 44.13: Surgical emphysema

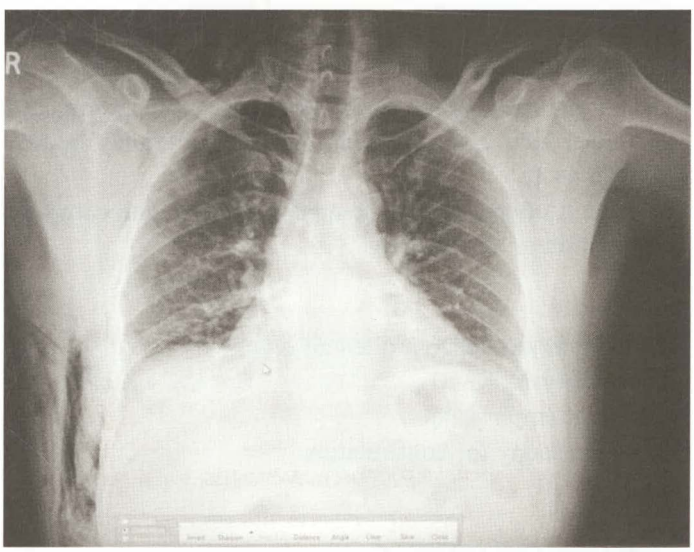


Fig. 44.14: Chest X-ray showing 10th rib fracture with surgical emphysema on the right side

KEY BOX 44.8

SURGICAL EMPHYSEMA

- Air in the subcutaneous tissue
- Palpable crepitus
- Sometimes, can be gross
- Disfigurement is more than symptoms and signs
- Rib stabbing the lung
- Rupture of bronchus—mediastinal emphysema

- The emphysema usually subsides within a week. Multiple incisions and expelling the air from the subcutaneous space manually are not required but may be carried out for cosmetic purposes or if patient has respiratory distress (Key Box 44.8).

PENETRATING THORACIC WOUNDS

Stab wounds

They depend on type of weapon, length and direction of the stab (Key Box 44.9).

Projectile wounds

- Relatively low velocity revolver bullets may perforate one or two lobes of the lung with a little damage and only drainage of the pleural space is required.
- High velocity perforating wounds cause more damage to the tissue, adjacent to the tract. If the damage is extensive, lobectomy or pneumonectomy has to be considered.
- Bomb fragments, because of irregular shape, commonly carry with them pieces of ribs and are associated not only with **severe haemorrhage** and air leak from the torn pulmonary vessels and bronchus but also with haemorrhage from irregular entry and exit wounds. Urgent thoracotomy is necessary.

Treatment

Emergency thoracotomy is a life-saving procedure in the trauma centre. Thoracotomy for polytrauma has a poor prognosis than for isolated thoracic injuries.

Indications

- Decompression of cardiac tamponade
- Control of bleeding, allow for internal cardiac massage
- Clamping of descending thoracic aorta for exsanguinous bleeding in the abdomen.

Surgery

Anterolateral thoracotomy through 5th intercostal space, entered in 1–2 minutes.

KEY BOX 44.9

PENETRATING WOUNDS

- Represents mainly stab and gunshot wounds
- Immediately pack the wound
- Intercostal chest tube
- Thoracotomy mandatory

MEDIASTINAL EMPHYSEMA

- The emphysema is mainly **suprasternal.**
- On auscultation, **pericardial crunching sounds** synchronous with the heart beat, are heard.
- If there is haemodynamic instability, bilateral intrapleural ICT has to be introduced as a precaution against tension pneumothorax.
- Rule out oesophageal and tracheal injury.

Principles of managing chest injuries

1. **Pulmonary physiotherapy:** Most important in all chest injury patients.
2. **Aspiration of secretions:** Tracheal aspiration, nasotracheal suction, aspiration of oral cavity and pharynx.
3. **Relieving pain:** Oral narcotics, parenteral narcotics, thoracic epidural analgesia, intercostal nerve block.
4. **Physiotherapy assistance**
   - Encourage coughing
   - Chest percussion and vibration
   - Deep inspiratory efforts
   - Humidification of air, nebulisation
   - Early mobilisation
5. **Treatment of pneumothorax or haemothorax:** Insertion of intercostal chest tube.
6. **Treatment of shock:** First of all, the causes of shock in chest injuries has to be determined by thorough assessment of the patient. Once the cause is found out, depending upon the nature of the problem, the patient is treated in an intensive care unit (Key Box 44.10).

KEY BOX 44.10

CAUSES OF SHOCK

- Tension pneumothorax
- Massive haemothorax
- Cardiac tamponade
- Myocardial contusion
- Air embolism
- Ruptured diaphragm
- Injury to the great vessels

7. **FAST (focussed abdominal sonography in trauma) ultrasound:** Focussed assessment with ultrasonography for trauma and rapid assessment for ruling out fluid collection in the abdomen, chest and pericardium.
8. **Surgery:** Depending upon the severity and location, surgery is done.
9. **Treatment of complications** (Key Box 44.11)
   - Thromboembolism
   - Tracheostomy complications
   - Prolonged ICU complications.

KEY BOX 44.11

COMPLICATIONS

- Empyema
- Bronchopleural fistula
- Bronchial stenosis
- Chylothorax
- Clotted haemothorax
- ARDS
- Atelectasis

Nontraumatic rib fractures (Key Box 44.12)

KEY BOX 44.12

NONTRAUMATIC RIB FRACTURES

- Stress fractures
- Metastatic disease
- Metabolic disease such as hyperparathyroidism
- Osteogenesis imperfecta
- Consider child abuse, 'shaken baby' syndrome
- Older patients after violent coughing

EMPYEMA

Definition

Collection of the pus in the pleural space is called empyema.

Aetiopathogenesis

- It is the end stage result of pleural effusion and infection. A few examples are—following haemothorax, lung abscess, pneumonia. In India, tuberculosis is an important cause of empyema (Figs 44.15 and 44.16)
- Oesophageal perforations—iatrogenic or spontaneous also result in empyema.
- Rupture of subphrenic abscess, rupture of hydatid cyst, rupture of amoebic liver abscess also can result in empyema.
- The classical events which follow empyema following pneumonia are exudative phase with pleural effusion, followed by thickening of fluid—fibropurulent stage and when the lung is covered by thick cortex, it is called organising phase.

Diagnosis

- History of fever diagnosed as pneumonia or tuberculosis
- Pain in the chest, difficulty in breathing
- Tenderness over the chest
- Toxic features in acute empyema specially in children
- Presence of thick pus with thick cortex of fibrin and coagulum over the lung.

Investigations

- Chest X-ray may show collapse of the lung, tracheal deviation, evidence of pneumonia or tuberculosis.
- Aspiration of pleural fluid and analysis—exudative in cases of pneumonia. Send for bacterial culture.
- CT scan may show split pleura sign. Tuberculous spine can be diagnosed as a cause of empyema (Figs 44.17 to 44.18)

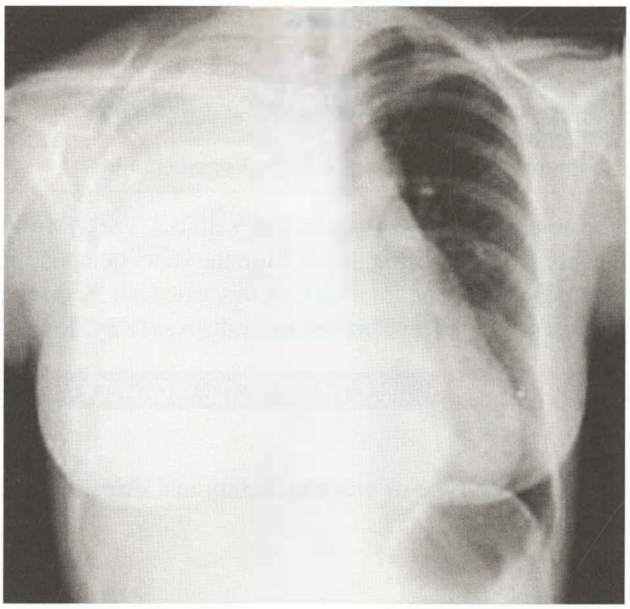


Fig. 44.15: Effusion

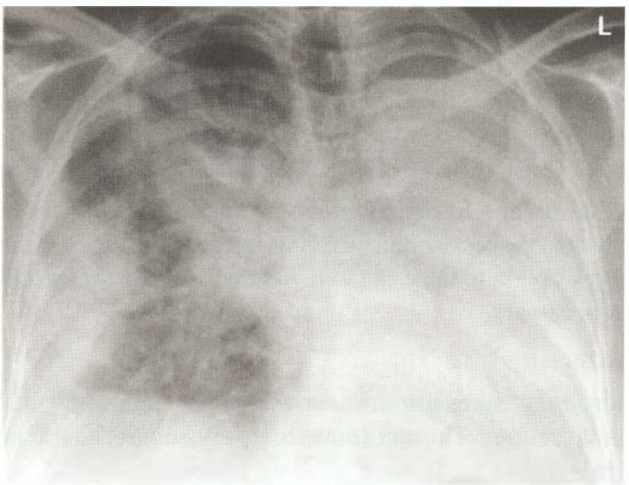


Fig. 44.16: Consolidation

Fig. 44.17: CT right lung empyema

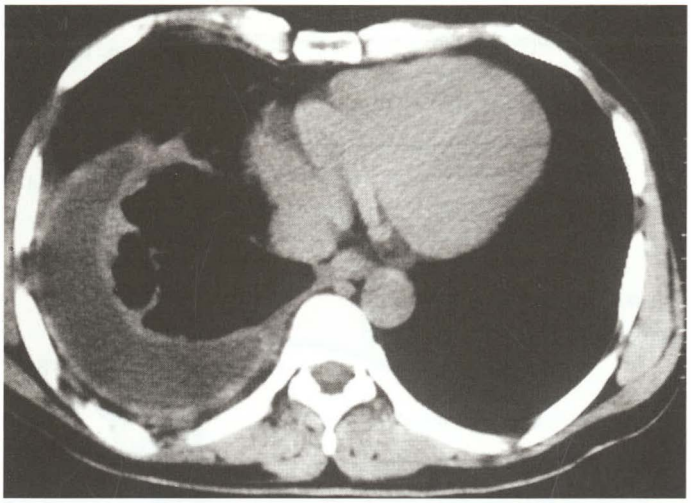


Fig. 44.18A: Right empyema split pleura sign

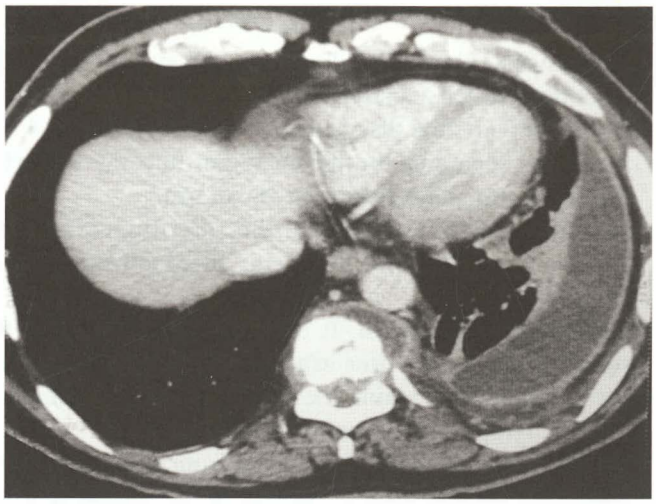


Fig. 44.18B: Empyema left with Koch's spine—split pleura sign

(*Courtesy:* Figs 44.15 to 44.18 are contributed by Dr C.S. Rajan, Senior Consultant and Head, Dept of Thoracic Surgery, St. Martha's Hospital, Bangalore)

Surgical management of pleural effusions and empyema

- **V**ideo **A**ssisted **T**horacoscopic **S**urgery (**VATS**): With 3–4 ports, video assisted procedures have become very popular. Minimal incision, less pain and recovery is fast. Drainage, pleural biopsy, talc pleurodesis, debridement of empyema, intercostal tube drainage (ICD) can all be done.
- Rib resections, and drainage through a window—Eloisier's method.
- **Decortication:** More radical procedure involves thoracotomy, debridement, excision of thick cortex or the covering of the lung, so that lung will expand. This is done by posterolateral thoracotomy.

Complications

- Toxicity, septic shock, multiorgan failure in untreated cases
- Damage to the lungs—lobectomy, pneumonectomy

Empyema necessitans: It is a type of empyema which presents as a swelling in the subcutaneous plane with communication to the pleural space/cavity. It is a tense, tender, fluctuant swelling with local rise of temperature. Intercostal bulge is also seen. On asking the patient to cough, expansile impulse is felt. Otherwise, management is similar to that of empyema such as drainage, treating the cause, ATT in tuberculosis and antibiotics in pyogenic infections.

BRONCHOPLEURAL FISTULA

Definition

It is a fistulous communication between the pleural space and the lung.

Causes

- Following pneumonectomy or an infection.
- It may occur when large airways are in communication with the pleural space following a large pneumothorax.

Pathogenesis

- There will be a large empty space left behind following pneumonectomy. This space will be filled with air. Over a period of time air is absorbed. The gaping of bronchial stump occurs resulting in bronchopleural fistula.
- Invariably the fluid which accumulates later will be infected—purulent.

Clinical features

- History and clinical features suggestive of empyema
- History of lung surgery
- Persistent air leak in the intercostal drain
- Pus in the ICD

Treatment is extremely difficult and disappointing

- Propped up/sitting position and turn to the disease side so as to get a dependent drainage
- Intercostal drain connected to under water seal
- Pleurocutaneous window drainage can be done
- Specific treatment includes control of infection, treat the primary cause, suturing, etc.

SURGICAL ANATOMY OF MEDIASTINUM AND MEDIASTINAL MASSES

The **mediastinum** is a broad central partition that separates the two laterally placed pleural cavities. It extends from the sternum to the bodies of the vertebrae; and from the superior thoracic aperture to the diaphragm.

Contents: The thymus gland, pericardial sac, heart, trachea and major arteries and veins. It also serves as a passageway for structures such as the oesophagus, thoracic duct and various components of the nervous system as they traverse the thorax on their way to the abdomen.

Division of mediastinum (Fig. 44.19)

- A transverse plane extending from the sternal angle (the junction between manubrium and the body of the sternum) to the intervertebral disc between vertebrae thoracic 4 and 5 spaces separates the mediastinum into the **superior mediastinum** and **inferior mediastinum**.
- **Inferior mediastinum** is further partitioned into the **anterior, middle, and posterior mediastinum** by the pericardial sac.
- **Anterior mediastinum**: It lies between the back of the sternum and the anterior aspect of the great vessels and pericardium. It contains the thymus, internal mammary arteries, lymph nodes, connective tissue and fat.
- **Middle mediastinum:** It extends from the pericardium anteriorly to the ventral surface of the thoracic spine posteriorly. It contains the pericardium, heart, great vessels, airway and oesophagus.
- **Posterior mediastinum:** It is made up of the spine and includes the costovertebral sulci. The posterior compartment contains the proximal intercostal neurovascular bundles, the spinal ganglia, the sympathetic chain, lymphatic tissue, and connective tissue.

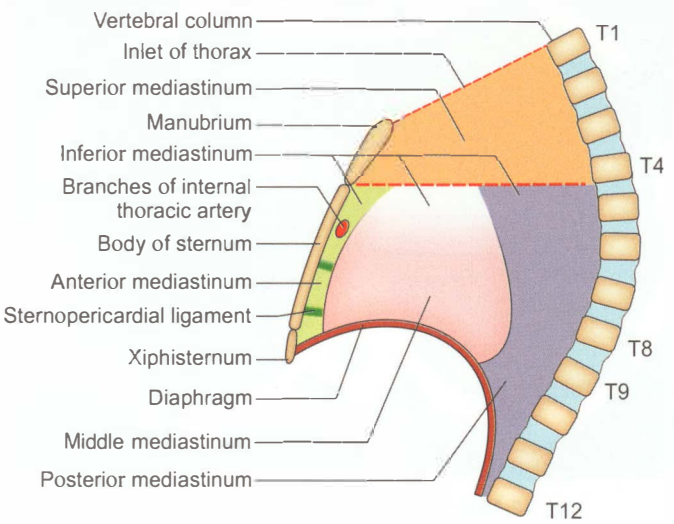


Fig. 44.19: Subdivisions of the mediastinum

Differential diagnosis of mediastinal masses is shown in Table 44.2.

Table 44.2 Differential diagnosis of mediastinal masses

Anterior mediastinum	Middle mediastinum	Posterior mediastinum
Thymus • Thymoma • Thymic cyst • Thymic hyperplasia • Thymic carcinoma	Bronchogenic cyst	**Neurogenic tumours** • Neurofibroma • Neurilemmoma • Neurosarcoma • Ganglioneuroma • Ganglioneuroblastoma • Neuroblastoma • Chemodectoma • Phaeochromocytoma
Lymphoma	Pericardial cyst	Meningocoeles
Germ cell tumour • Teratoma/dermoid cyst • Seminoma • Nonseminoma – Yolk sac tumour – Embryonal carcinoma – Choriocarcinoma	Lymphadenopathy • Lymphoma • Sarcoid • Metastatic lung cancer	Thoracic spine lesion (e.g. Pott's disease)
Intrathoracic thyroid • Substernal goitre • Ectopic thyroid tissue	Enteric cyst	
Parathyroid adenoma	Oesophageal tumours	
Haemangioma	Vascular masses and enlargement	
Lipoma		
Liposarcoma		
Fibroma		
Fibrosarcoma		
Foramen of Morgagni hernia		

ANTERIOR MEDIASTINAL MASSES

"Terrible Ts": Thymic tumours, Teratoma/germ cell tumour, (Terrible) lymphoma, and Thyroid.

1. Thymoma

- Most common anterior mediastinal primary tumour; 20% of adult mediastinal neoplasms.
- Presentation between ages 30 and 50 (most patients are > 40 years old).
- 50% are asymptomatic but others have symptoms secondary to compression: Chest pain, cough, dyspnoea, compression of the superior vena cava resulting in head and neck venous congestion, facial oedema.
- **Myasthenia gravis:** Seen in 30–50% of thymoma patients; others can have hypogammaglobulinemia (10%), endocrine disorders, connective tissue disorders.

Staging (Table 44.3)

Investigations

- Chest X-ray may show a mediastinal mass.
- Contrast CT scan of the thorax is the investigation of choice which will detect a mass lesion and its relationship to the adjacent structures (Fig. 44.20).
- MRI can also be done
- Preoperative biopsy is not necessary (Key Box 44.13).

Surgery

- Surgery is the main modality of the treatment—a complete resection of the thymus. Median sternotomy is done to remove the tumour.

Table 44.3 Masaoka's clinical stage

Stage I:	Macroscopically completely encapsulated and microscopically no capsular invasion
Stage II:	Macroscopic invasion into surrounding fatty tissue or mediastinal pleura, or microscopic invasion into capsule
Stage IIII:	Macroscopic invasion into neighbouring organ, i.e. pericardium, great vessels, or lung
Stage IVa:	Pleural or pericardial dissemination
Stage IVb:	Lymphogenous or haematogenous metastasis

KEY BOX 44.13

PREOPERATIVE BIOPSY IS NOT NECESSARY FOR THE RESECTION OF FOLLOWING TUMOURS

1. Thymoma
2. Pleomorphic adenoma
3. Testicular tumours
4. Renal cell carcinoma
5. Hepatocellular carcinoma

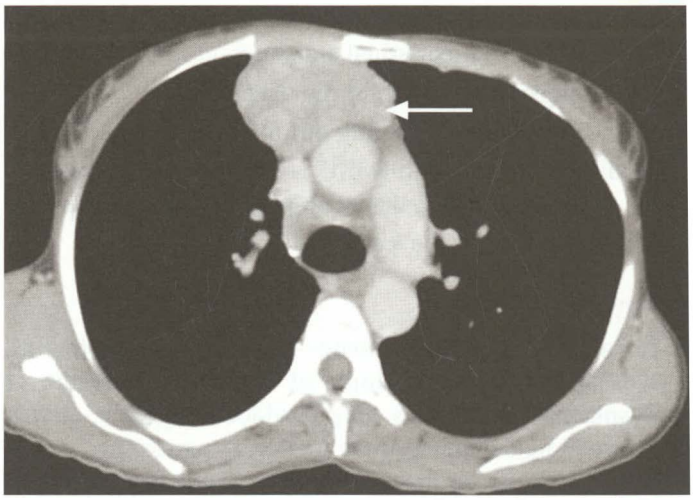


Fig. 44.20: Thymoma

- A minimally invasive (thoracoscopic or robotic) approach is another option.

2. Germ cell tumours (GCT)

- The mediastinum is the most common location for extragonadal germ cell tumours in adults.
- They are classified as benign (teratomas, dermoid cysts) or malignant (seminomas, nonseminomatous GCTs).
- Seminomas are more common than nonseminomatous GCTs.
- Occur in the second decade—late teens
- 20% are malignant
- Cystic teratomas are dermoid cysts, less malignant compared to solid teratomas which have more chances of malignancy.
- Diagnosis is by tumour markers: Alfa fetoproteins (AFP) and beta-hCG. AFP is normal in teratoma and "pure" seminomas. Ninety percent of nonseminomatous germ cell tumours have elevated AFP and/or beta-hCG.
- They are highly radiosensitive (Key Box 44.14).

KEY BOX 44.14

TUMOURS WHICH ARE HIGHLY RADIOSENSITIVE

1. Germ cell tumours
2. Seminoma
3. Lymphoma
4. Squamous cell carcinoma

3. Lymphomas

- The most common are nodular sclerosing Hodgkin's lymphoma and primary mediastinal B-cell lymphoma.
- May present with systemic symptoms such as fevers, weight loss or night sweats but can also present with symptoms such as chest pain, dyspnoea, wheezing, stridor, hoarseness,

dysphagia, or **superior vena cava syndrome** due to compression of mediastinal structures.

- **Core biopsy** is required for the diagnosis
- **CT scan will help in ruling out other diseases**.
- Mediastinal mass ratio (MMR)—the ratio of the mass to the chest diameter is calculated (*see* page 119 of Chapter 9)
- **Chemotherapy** is the treatment of choice.

4. Thyroid masses

- Intrathoracic thyroid tissue—**ectopic or substernal**, typically causes symptoms of shortness of breath or dysphagia. Sometimes they produce dangerous airway obstruction, deviation of the trachea or narrowing.
- The intrathoracic mass is usually continuous with the thyroid gland in the neck; only 2 percent of cases are separate from the cervical thyroid and are truly intrathoracic.
- Diagnosis is by CT scan and technetium scan.
- Majority can be managed with cervical incision—a few may require sternotomy.
- Less commonly, intrathoracic mobilisation *via* **VATS** is done.
- Possibility of collapse of trachea should be kept in mind after thyroidectomy—one should be ready with re-intubation/tracheostomy.

MIDDLE MEDIASTINAL MASSES

- Lymphadenopathy is the most common lesion presenting as a mass in the middle compartment of the mediastinum.
- The most common causes include lymphoma, sarcoid and metastatic lung cancer.
- Mediastinoscopy is a very useful technique to biopsy lymphadenopathy in this region.
- Cystic masses comprise approximately 20 percent of middle mediastinal masses.

1. Bronchogenic cysts

- Bronchogenic cysts are the most common cystic lesion
- Secondary to abnormal lung budding during development.
- Bronchogenic cysts are more common in men, in the right paratracheal region and in the subcarinal location.
- Often presents with substernal pain, cough, recurrent infection symptoms or dyspnoea.

2. Enteric cysts

- They are the third most common benign oesophageal masses after leiomyomas and polyps, and are usually asymptomatic.
- **Three criteria** are required to establish their diagnosis:
  (a) Oesophageal attachment
  (b) The presence of two layers of muscularis propria
  (c) Epithelium characteristic of the gastrointestinal tract.

3. Pericardial cysts

- Seventy percent arise in the right cardiophrenic angle.
- Symptoms can include shortness of breath, right heart failure secondary to compression, infection and bleeding.

Management

- Bronchogenic and enteric cysts need surgical resection to establish a definitive diagnosis and to decrease the risk of infection or malignant degeneration.
- Pericardial cysts can typically be observed if asymptomatic but resection can be utilised if there are symptoms or if the diagnosis is not completely established by imaging.
- Simple drainage is generally not recommended because *these cysts typically* recur without complete resection.

POSTERIOR MEDIASTINAL MASSES

Neurogenic tumours represent more than 60 percent of posterior mediastinal masses. These lesions are classified based upon their neural cell of origin. 95% of posterior mediastinal masses arise in the intercostal nerve rami or the sympathetic chain region.

1. Schwannomas and neurofibromas

- These are benign lesions that arise from the intercostal nerve sheath and make up 90 percent of adult neurogenic tumours.
- Neurilemmomas or schwannomas constitute 75% of this group of masses. These tumours are firm, encapsulated masses consisting of Schwann cells.
- Neurofibromas are nonencapsulated, soft, friable and are associated with von Recklinghausen neurofibromatosis.
- The surgery of choice for removal of these tumours is by thorocoscopy or thoracotomy.
- Postoperative complications include Horner's syndrome, partial sympathectomy, recurrent laryngeal nerve damage and paraplegia.

2. Malignant tumours of nerve sheath origin

- Malignant nerve sheath tumours are spindle cell sarcomas of the posterior mediastinum and include malignant neurofibromas, malignant schwannomas and neurogenic fibrosarcomas.
- They affect men and women equally in the third to fifth decade of life and are closely associated with neuro-fibromatosis, with a 5% risk of sarcomatous degeneration.
- Pain and nerve deficits are common.
- Complete surgical resection is the optimal treatment but in patients with unresectable tumours, adjuvant chemotherapy and radiation are options.

3. Autonomic ganglionic tumours

- Neuroblastomas and ganglioneuroblastomas are malignant tumours that occur most commonly in children and originate from the sympathetic ganglia.

- Ganglioneuromas are benign lesions that arise from the sympathetic ganglia, and are most common in young adults. Lesions that arise from paraganglionic cells include phaeochromocytomas and paragangliomas.
- Some neurogenic tumours are "dumb-bell shaped" and arise near intervertebral foramen, and have a posterior mediastinal and intraspinal component.
- Resection usually requires a combined approach with neurosurgery and thoracic surgery.

PULMONARY ASPERGILLOMA

- Pulmonary **aspergilloma**, is a **mycetoma** or **fungus ball**, caused by fungus of *Aspergillus* species
- The most common place affected by aspergillomas is the lung
- Tuberculosis of the lungs and immunocompromised conditions are risk factors.
- The fungus settles in a cavity and grows to a big ball—called fungal ball
- Majority of the cases are asymptomatic
- Cough, chest pain, haemoptysis, abscess formation are the features
- Diagnosis is by chest X-ray and CT scan (Fig. 44.21), majority of the cases do not require any treatment
- In symptomatic cases (rarely), removal of the lesion, lobectomy may be required (Fig. 44.22).

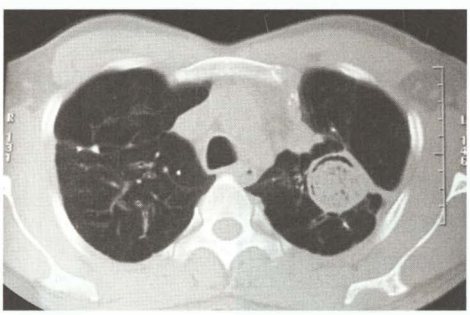


Fig. 44.21: CT chest showing crescent sign—pulmonary aspergilloma

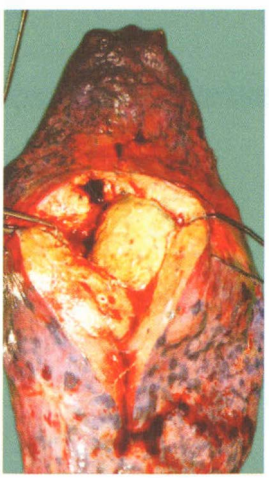


Fig. 44.22: Pulmonary aspergilloma—cut open specimen
(*Courtesy:* Figs 44.21 and 44.22, Dr C.S. Rajan, Senior Consultant and Head, Dept of Thoracic Surgery, St. Martha's Hospital, Bangalore)

BRONCHOGENIC CARCINOMA

Introduction

Bronchogenic cancer is the most frequent cause of cancer death in men and women and accounts for 14.5% of all cancer diagnoses and 27.6% of all cancer deaths in the United States. Cigarette smoking is unequivocally the most important risk factor in the development of lung cancer. In India, bronchogenic carcinoma has become number one cancer in men overtaking other cancers. The diagnosis is often delayed. Many patients present as metastasis in bones. Surgical resection alone cannot be curative in many cases because of late presentation of the cases. Prognosis is poor in spite of the treatment.

Risk factors

- **Cigarette smoking:** It is described as pack years—a pack containing 20 cigarettes.
- **Industrial carcinogens:** Asbestos, arsenic, chromium or nickel, organic chemicals, radon.
- **Radon** is the second leading cause of lung cancer. Radon is a natural radioactive gas released from the normal decay of uranium in the soil.

Pathology—types

1. **Adenocarcinoma** (ADA) of the lung is the most frequent histologic type and accounts for approximately 45% of all lung cancers. Women smokers have more incidence of adenocarcinoma. Adenocarcinoma of the lung develops from the mucus-producing cells of the bronchial epithelium. Cells are cuboidal to columnar cells. Most of these tumours (75%) are peripherally located. Adenocarcinoma of the lung tends to metastasise earlier than squamous cell carcinoma of the lung and more frequently to the central nervous system (CNS)—cases can present as weakness of the limbs, hemiparesis, etc.
2. **Bronchioloalveolar carcinoma** is an adenocarcinoma. Sometimes it can be a more indolent disease. It is well differentiated and spreads along alveolar walls without invasion of stroma, blood vessels or pleura.
3. **Squamous cell carcinoma** (SCCA) of the lung occurs in approximately 30% of patients with lung cancer. Approximately 75% of these tumours are centrally located and tend to expand against the bronchus, causing extrinsic compression. These tumours are prone to undergo central necrosis and cavitation. **Squamous cell carcinoma** tends to metastasise later than adenocarcinoma. Microscopically, keratinisation, stratification and intercellular bridge formation are exhibited. SCCA may be more readily detected on sputum cytology than ADA.
4. **Large cell undifferentiated carcinoma** may be made in approximately 10% of all lung tumours. These tumours tend to occur peripherally and may metastasise relatively early. Small cell lung cancer represents approximately 20% of

Table 44.4 Differences between adenocarcinoma and squamous cell carcinoma

Factors	Adenocarcinoma	Squamous cell carcinoma
1. Incidence	Most common—45%	30%
2. Metastasis	Early	Later
3. Location	Peripheral	Central
4. Metastasis to brain	More frequent	Less frequent

all lung cancers; approximately 80% are centrally located. The disease is characterised by an aggressive tendency to metastasise. It often spreads early to mediastinal lymph nodes and distant sites, especially bone marrow and brain.

5. **Small cell lung cancer** appears to arise in cells derived from the embryologic neural crest. Microscopically, these cells appear as **sheets or clusters of small dense cells**, with dark nuclei and little cytoplasm. This oat-like appearance under the microscope gives the term oat cell carcinoma to this disease. These tumours are often advanced at presentation, with an aggressive tendency to metastasise both by lymphatics and by blood. Chemoradiotherapy is generally used for treatment. However, the 5-year survival rate is only 5%.

Table 44.4 shows differences between adenocarcinoma and squamous cell carcinoma.

Clinical features

- Cough and haemoptysis: It is a nonspecific symptom in many patients and diagnosis is delayed because it is often thought to be a smoker's cough.
- Dyspnoea is due to pleural effusion or due to restrictive pulmonary disease.
- Bloody effusion, clubbing of the fingers, localised chest pain are other features.
- Hoarseness due to recurrent laryngeal nerve paralysis, back ache due to metastasis in vertebrae, or neurological features due to metastasis in brain are also presenting features in many cases.
- Myopathy similar to myasthenia gravis can occur in small cell carcinoma.

Diagnosis and spread

- In addition to chest X-ray, CT scan, sputum for malignant cells, diagnosis can also be made with bronchoscopic biopsy and CT-guided biopsy. Spread is common PET scan: 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) is the investigation of choice to detect the distant lesions (Figs 44.23 to 44.25).
- Brain CT or magnetic resonance imaging (MRI) (Fig. 44.26)
- Mediastinoscopy

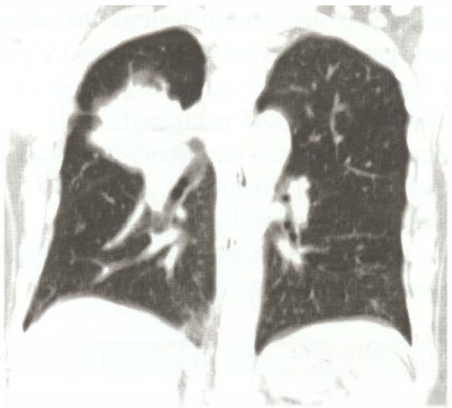
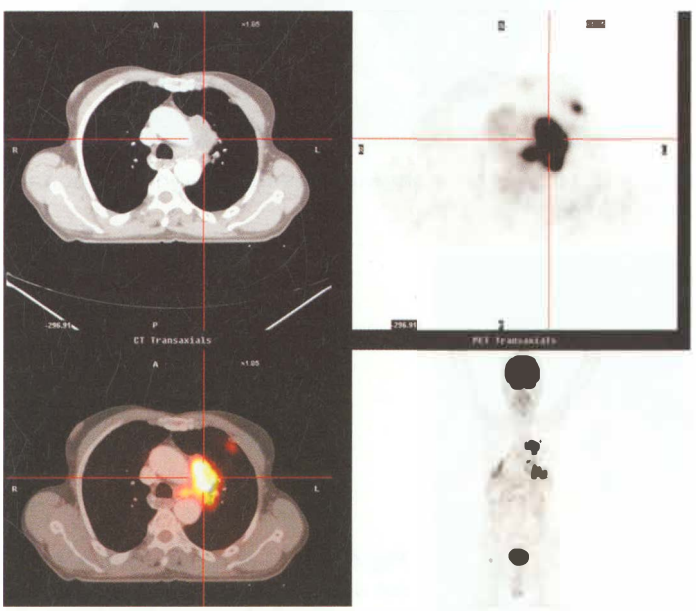


Fig. 44.23: CT of lungs showing bronchogenic carcinoma

Fig. 44.24: Sagittal section of the CT showing bronchogenic carcinoma right lung



PET-CT of a patient who was diagnosed to have carcinoma lung on bronchoscopy.
PET CT shows a hilar mass with a nodule anteriorly on left side of pleura.
It also shows pneumonic patch on lower zone of left lung which is FDG avid

Fig. 44.25: PET scan of lung showing bronchogenic carcinoma

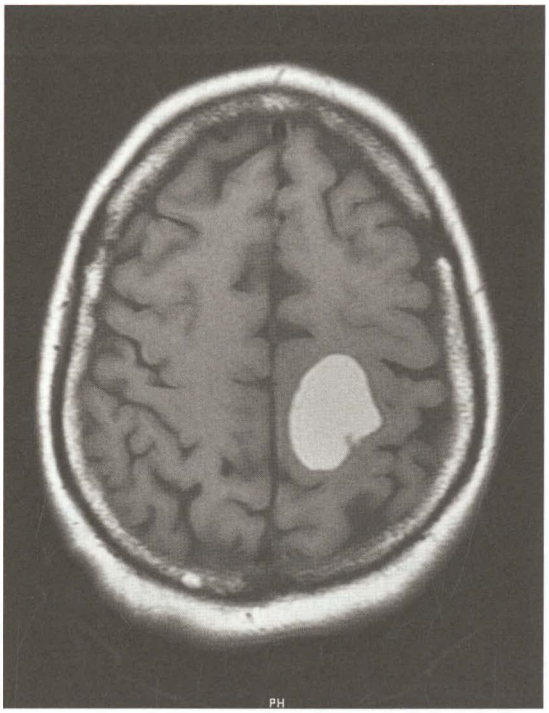


Fig. 44.26: MRI brain showing metastasis from bronchogenic carcinoma

TNM STAGING

Stage	T	N	M
Occult cancer	TX	N0	M0
0	Tis	N0	M0
IA	T1a/b	N0	M0
IB	T2a	N0	M0
IIA	T2b	N0	M0
	T1a/b; T2a	N1	M0
IIB	T2b	N1	M0
	T3	N0	M0
IIIA	Any T1; T2	N2	M0
	T3	N1/N2	M0
	T4	N0/N1	M0
IIIB	T4	N2	M0
	Any T	N2	M0
	Any T	N5	M0
IV	Any T	Any N	M1a/b

- CT of the chest, abdomen; MRI brain: Mediastinal lymph node masses, metastases to the adrenal glands, brain, lung and bone are common.
- Bone metastases are osteolytic. Whole body bone scan is needed.

TNM STAGING The international tumour—node-metastasis

T (Primary tumour)

TX: Primary tumour cannot be assessed, or tumour proven by the presence of malignant cells in sputum or bronchial washings but not visualised by imaging or bronchoscopy

T0: No evidence of primary tumour
Tis: Carcinoma *in situ*
T1: Tumour ≤ 3 cm in greatest dimension, surrounded by lung or visceral pleura, without bronchoscopic evidence of invasion, more proximal than the lobar bronchus (i.e. not in the main bronchus)
- T 1a tumour ≤ 2 cm in greatest dimension
- T 1b tumour > 2 cm but ≤ 3 cm in greatest dimension

T2 tumour > 3 cm but ≤ 7 cm or tumour with any of the following features:
- Involves main bronchus ≥ 2 cm distal to the carina
- Invades visceral pleura associated with atelectasis or obstructive pneumonitis that extends to the hilar region but does not involve the entire lung
- T2a tumour > 3 cm but ≤ 5 cm in greatest dimension
- T2b tumour > 5 cm but ≤ 7 cm in greatest dimension

T3 tumour > 7 cm or one that directly invades any of the following: chest wall (including superior sulcus tumours), diaphragm, phrenic nerve, mediastinal pleura, parietal pericardium; or tumour in the main bronchus < 2 cm distal to the carina but without involvement of the carina or associated atelectasis/obstructive pneumonitis of the entire lung or separate tumour nodule(s) in the same lobe

T4 tumour of any size that invades any of the following: Mediastinum, heart, great vessels, trachea, recurrent laryngeal nerve, oesophagus, vertebral body, carina; or separate tumour nodule(s) in a different ipsilateral lobe

N (Regional lymph nodes)

NX: Regional lymph nodes cannot be assessed
N0: No regional lymph node metastasis
N1: Metastasis in ipsilateral peribronchial and/or ipsilateral hilar lymph nodes and intrapulmonary nodes including involvement by direct extension
N2: Metastasis in ipsilateral mediastinal and/or subcarinal lymph node(s)
N3: Metastasis in contralateral mediastinal, contralateral hilar, ipsilateral or contralateral scalene or supraclavicular lymph node(s)

M (Distant metastasis)

MX: Distant metastasis cannot be assessed
M0: No distant metastasis
M1: Distant metastasis
- M1a separate tumour nodule(s) in a contralateral lobe; tumour with pleural nodules or malignant pleural (or pericardial) effusion
- M1b: Distant metastasis

Treatment

Can be grouped into three major categories

1. Stages I and II tumours are contained within the lung and may be completely resected with surgery.
2. Resectable stages IIIA and IIIB tumours are locally advanced tumours with metastasis to the ipsilateral mediastinal (N2) lymph nodes (stage IIIA) or involving mediastinal structures (T4N0M0). These tumours, by their advanced nature, may be mechanically removed with surgery followed by systemic chemotherapy. Radiotherapy can also be given to locally advanced disease.
3. Stage IV disease includes metastatic disease and is not typically treated by surgery, except for patients requiring surgical palliation. Systemic therapies—chemotherapy for metastatic disease are common.

CONGENITAL HEART DISEASES

Introduction

- Abnormal heart structures since birth.
- It develops between 3rd and 8th week of foetal life.
- 1st operation for congenital heart disease was ligation of PDA by Cross in 1938.
- With new advances in neonatal cardiopulmonary bypass (CPB), outcomes have improved.

Development

- At 12 weeks, primitive vascular tube is fully developed.
- Foetal circulation differs from adult in that right and left ventricles pump blood in parallel rather than in series to receive increasingly oxygenated blood.
- **3 important structures are ductus venosus, foramen ovale and ductus arteriosus.**

Changes at birth

- Pulmonary vascular resistance falls (breathing)
- Pulmonary vasodilation and ductus arteriosus constricts within 30 minutes of delivery (due to increased oxygen levels).
- Reversal of pulmonary and systemic pressure gradient. Termination of blood flow from pulmonary artery into aorta.
- Taping/cutting umbilical cord
- Venous blood from placenta stops
- IVC and right atrial pressure decrease and left right atrial pressure increases (due to increased systemic vascular resistance)
- Foramen ovale closes

Abnormalities

- Persistence of normal channels (patent ductus arteriosus—PDA, patent foramen ovale).
- Failure of septation (atrial septal defect—ASD, ventricular septal defect—VSD, tetralogy of Fallot—TOF).
- Stenosis (intracardiac—supravalvular, valvular, infravalvular, coarctation of aorta).
- Atresia/abnormal connections (transposition of great arteries—TGA, total anomalous pulmonary venous connections—TAPVC).

Incidence

Eight cases per 1000 live births in UK

Aetiology

Unclear: Infection/Envirotoxins/Genetics

Diagnosis

In utero foetal ECHO

Classification

- Cyanotic (1/3) and acyanotic (2/3)
- Right to left shunts (similar to TOF)
- Left to right shunts (similar to ASD, VSD, PDA)
- Parallel systemic and pulmonary flow (similar to TGV)
- Aortic stenosis, coarctation of aorta
- Mixing of systemic and pulmonary flow (similar to TAPVC): Heart failure in infancy.

CYANOTIC CONGENITAL HEART DISEASE

1. Tetralogy of Fallot (TOF)

Most common cyanotic congenital heart disease

Child adopts squatting posture during hypoxic spells which increases systemic vascular resistance and venous return to heart. This directs the blood into pulmonary circulation and oxygenation improves. Chest X-ray shows boot-shaped heart. Treatment—single or two stage surgery (Key Box 44.15).

KEY BOX 44.15

COMPONENTS

1. VSD
2. Overriding aorta
3. Pulmonary stenosis
4. Right ventricular hypertrophy

2. Transposition of great vessels (TGA)

- **Most common cause of cyanosis in newborn.**

Features

- Aorta arises from right ventricle and pulmonary artery from left ventricle.
- Incompatible with life unless associated with ASD or VSD.
- **Chest X-ray—'Egg on side' appearance.**

Treatment

- Rashkind percutaneous balloon atrial septoplasty.
- Definitive repair—arterial switch (2 stage)
- Mustard or Senning surgery.

3. Total anomalous pulmonary venous drainage

- Pulmonary venous drain disconnected from left atrium and drains to systemic circulation, e.g. inferior vena cava (IVC) or superior vena cava (SVC).
- Presents with failure to thrive.
- Treatment—surgery

4. Eisenmenger's syndrome

- It occurs following reversal of a previous left to right shunt as with ASD or VSD.
- Similar to pulmonary hypertension.
- Closure of shunt contraindicated.

ACYANOTIC CONGENITAL HEART DISEASE

1. Patent ductus arteriosus

Definition

Persistence of the foetal ductus arteriosus in the postnatal period.

Embryology

- Derived from the 6th aortic arch
- Essential for the foetal circulation
- Blood ejected by right ventricle flows exclusively through the ductus to the lower extremity and placenta bypassing the high resistance pulmonary circulation in the foetus.

At birth

- Physiological closure occurs in 1–6 days
- Anatomical closure occurs in 2–3 weeks

Mode of closure

Smooth muscle constriction in response to rising arterial oxygen tension.

Pathophysiology

Shunt occurs across the ductus both in systole and diastole resulting in left ventricle overload and pulmonary plethora.

- Shunt depends on the size of the ductus and pulmonary and systemic vascular resistance. Continuous murmur is due to shunt during both systole and diastole.

Clinical features

- Incidence: M : F = 1:2
- It depends on the size of the ductus, pulmonary vascular resistance, age at presentation and associated anomalies.
  - **a. Small ductus: Asymptomatic**
  - **b. Infants:** Congestive heart failure
  - **c. Children:** Dyspnoea on exertion, repeated respiratory tract infection (Key Box 44.16)

KEY BOX 44.16

DIAGNOSIS

- Hyperdynamic precordium and bounding peripheral pulses in large shunt
- Machinery or **continuous murmur (Gibson's murmur)** in the left second intercostal space, radiating to the left infraclavicular area
- ECG: Left ventricular hypertrophy
- Chest X-ray: Cardiomegaly
- Pulmonary congestion in large ductus and hilar dance on fluoroscopy
- Two-dimensional echo is diagnostic—in suprasternal view, the PDA is seen

PEARLS OF WISDOM

Occurs 1 in 5000 live births. 50% in premature babies

Complications

- Reversal of shunt
- Infective endocarditis
- Congestive heart failure

Treatment

It can be divided into surgical and nonsurgical methods.

I. Surgical methods

- Presence of PDA is sufficient indication for surgery even without symptoms.
- **In infants:** Large PDA with congestive cardiac failure not responding to antifailure measures, surgery is indicated.

Surgery

- Triple ligation
- Division and suturing
- Right lateral position and posterolateral thoracotomy
- Thorax entered through 4th intercostal space
- **It can also be done by thoracoscopy** and video-assisted thoracoscopic surgery (VATS) and clipping of PDA.

Care during surgery

Left recurrent laryngeal nerve must be carefully preserved

Complications during surgery

- Haemorrhage, left recurrent laryngeal nerve palsy
- Chylothorax

II. Nonsurgical methods

- **Pharmacological closure:** In preterm infants—indomethacin
- **Transcatheter closure:** Interventional cardiology
- Double umbrella device—Rashkind
- Gianturco coils
- Lock Clamshell occluder.

2. Coarctation of aorta

Definition

Congenital narrowing of the descending thoracic aorta usually occurring just distal to the left subclavian artery origin, adjacent to the site of insertion of the ductus arteriosus.

Incidence

- 0.2–0.6 per 1000 live births
- 5–8% of all cases of congenital heart diseases.

Aetiology

Flow theory: Reduced flow in the aorta due to multiple abnormalities in the heart during the foetal period.

Ductal sling theory: When there is isolated coarctation, this theory is applicable. Abnormal extension of contractile ductal tissue into the adjacent aorta which results in coarctation.

Types

Infantile—preductal and adult—juxtaductal.

Classification

- Group I: Isolated coarctation
- Group II: Coarctation with VSD
- Group III: Coarctation with complex intracardiac anomalies (Key Box 44.17).

Clinical features

Depend on: Age and symptoms at presentation, location of the coarctation, severity of the coarctation, associated anomalies.

Symptoms

- Visual disturbances, exertional dyspnoea
- Upper extremity hypertension
- Headache, epistaxis
- Claudication of the lower limbs

Signs

- Systolic murmur heard over the precordium and posteriorly between the scapula.
- **Feeble femoral pulses.**
- Enlarged collateral vessels are seen (Suzman's sign) and palpable between scapula and bruit can be heard here.
- Systolic gradient between arm and leg blood pressure.

Collaterals

- Subclavian artery and its branches
- Internal mammary artery
- Intercostal artery
- Scapular, cervical, vertebral, epigastric and spinal arteries.

Investigations

- **ECG:** LVH with LV strain

KEY BOX 44.17

ASSOCIATED CONGENITAL ANOMALIES

- Ventricular septal defect
- Bicuspid aortic valve
- Patent ductus arteriosus
- Mitral valve abnormalities

- **CXR:** Notching of the ribs **(Dock's sign)** from third rib onwards, above four years of age.

Diagnosis

- Two-dimensional echocardiogram, cardiac catheterisation
- Transoesophageal echocardiogram (TEE)
- Angiogram

PEARLS OF WISDOM

Classic 'three' sign: Formed by dilated left subclavian artery, narrowing of the coarctation and the poststenotic dilatation of aorta.

Complications (Key Box 44.18)

KEY BOX 44.18

COMPLICATIONS

- Circle of Willis' aneurysm rupture
- Aortic dissection
- Early coronary atherosclerosis
- Bacterial endocarditis
- Congestive heart failure
- Spontaneous rupture of aorta

Treatment

A. Nonsurgical (infants)

B. Surgery

- Left posterolateral thoracotomy
- Thorax entered through fourth intercostal space
- Arterial line in the right upper limb
- Preserve the vagus and recurrent laryngeal nerves
- Arterial line in the lower limb
- Lower limb pressure during cross-clamping of aorta, to be kept above 45 mmHg
- If the pressure is low, then create a shunt from the aortic arch to the descending thoracic aorta with the help of a cannula.

Complications of surgery

- Haemorrhage, RLN injury, phrenic nerve injury
- Horner's syndrome, chylothorax
- Paradoxical hypertension, paraplegia, stroke.

Late complications

- Recoarctation, aneurysm and left arm ischaemia (when subclavian flap aortoplasty is done).

Pseudocoarctation

Rare condition which results from congenital elongation of the aortic arch which leads to redundancy and kinking of the aorta and may appear similar to coarctation but there is no obstruction to blood flow.

3. Atrial septal defect

Defect in septum between left and right atrium leading to left to right shunt.

Types: 3 types

- **Ostium secundum:** Defect in floor of fossa ovalis. Presents late in life.
- **Ostium primum:** Endocardial cushion defect associated with mitral valve defects (mitral regurgitation) and trisomy 21. Presents earlier in life.
- **Sinus venosus:** Defect near junction of SVC and atrium.

Treatment

Open heart surgery with CPB and closure of defect with sutures or with pericardial or synthetic patch.

4. Ventricular septal defect

Types: 4 types

- Perimembranous defect (in membranous septum): 70–80%
- Muscular defect (multiple): 10%
- Atrioventricular defect: 5%
- Subarterial defect: 5–10%

O/E: Pansystolic murmur

Treatment: Surgical repair

CORONARY ARTERY BYPASS SURGERY

Introduction

Often simply called bypass surgery, it is the **most commonly performed open heart surgery all over the world.** The incidence of coronary artery disease is rapidly rising in the developing countries. A **vein graft** from the lower limb or an **arterial graft** (internal mammary artery or the radial artery) is used to bypass the obstructed coronary artery. The bypass is done from the root of the aorta to the distal coronary artery.

- The obstruction or the block is not touched.

Risk factors for coronary artery disease (Key Box 44.19)

Incidence

Least in Japan and highest in Finland.

KEY BOX 44.19

RISK FACTORS

I. Nonmodifiable

- Age, sex, family history

II. Modifiable, controlled or treated by medicines:

- Smoking, uncontrolled diabetes
- Uncontrolled hypertension
- Obesity, overweight, lack of physical exercise
- Individual response to stress
- High blood cholesterol or lipid abnormalities

Indications for surgery

1. Usually indicated in symptomatic or for prognostic reasons (i.e. balance between expectant benefit and risk faced by patient).
   a. > 50% stenosis of left main stem artery
   b. > 70% stenosis of proximal left anterior interventricular artery
   c. Triple vessel disease
   d. Poor ventricular function associated with CAD.
2. Chronic stable angina
3. Acute coronary syndrome
4. Surgery for complications of MI such as ventricular septal rupture, etc.

PEARLS OF WISDOM

With interventional cardiology progressing rapidly the indications for surgery are getting blurred.

Investigations

- ECG/cardiac enzymes—Trop T and Trop I
- 2D echocardiogram—new stress ECHO
- Holter monitoring, thallium scan-201 or ^{99m}Tc
- Stress test (exercise tolerance test)
- Coronary angiogram
- Radionucleotide studies and cardiac MRI
- CT (multislice high resolution)

Contraindications of CABG

- Small, diffusely diseased arteries
- Diffuse disease and heart failure
- Acute myocardial infarction over 6 hours old
- Moribund patients after resuscitation

Surgery

- Bypass surgery with vein or artery graft
- Endarterectomy
- Venous patch
- Transmyocardial laser revascularisation (TMLR).

Bypass surgery

1. Techniques of bypass

- **Cold cardioplegia** with moderate hypothermia and cardiopulmonary bypass.
- Ischaemic cross-clamp with moderate hypothermia with cardiopulmonary bypass.
- **Empty beating heart**, normothermic with cardiopulmonary bypass.
- **Beating heart surgery,** no heart lung machine.

2. Grafts

- Veins—lower limbs (long saphenous vein)
- Upper limb (cephalic vein) rarely used
- Arteries—radial (Key Box 44.20)
- Internal mammary artery both right and left
- Gastroepiploic
- Inferior epigastric
- Inferior mesenteric

KEY BOX 44.20

ADVANTAGES OF ARTERIAL GRAFT

- It is artery to artery anastomosis
- Compatible size
- Flow adaptation
- IMA advantages: No vasa vasorum
- Dense nonfenestrated intact internal elastic lamina that inhibits cellular migration and subsequent initiation of hyperplasia
- Thin medial layer with a few smooth muscle cells which provide a little vasoreactivity
- Produces more prostacyclines which is a vasodilator and platelet inhibitor

3. Nonautogenic conduits

- Cryopreserved human saphenous vein allograft
- Processed bovine sacral artery
- Polytetrafluoroethylene graft (Gore-Tex)

Standard surgical procedure

- Median sternotomy
- Internal mammary artery dissection done
- Pericardium opened
- Systemic heparinisation 3 mg/kg
- Aortic and venous cannulation.
- Cardiopulmonary bypass started
- Aorta cross-clamped and cold K^+ rich solution given in the aortic root
- Heart arrested in diastole to achieve immobile surgical field
- Distal anastomosis completed
- Proximal anastomosis done with the heart beating
- Weaned off CPB
- Protamine given and decannulation carried out
- Sternum closed, extubation 6–8 hours later.

Complications of surgery

1. Perioperative MI (2–3%)
2. Stroke
3. Arrhythmias (30%)
4. Sinus tachycardia
5. Atrial fibrillation
6. Reinfarct
7. Wound infection
8. Bleeding
9. General weakness
10. Mortality (2–3%)
11. Persistent poor cardiac output requiring inotropes o mechanical support

Mechanical support to low cardiac output patients post CABG: Intra-aortic balloon pump (IABP)

- Inserted percutaneously in common femoral artery, threade into aorta till its tip lies in distal arch vessels.
- Balloon is triggered by the ECG, deflating during ventricula systole (decreasing afterload) and inflating during diastol (increasing diastolic pressure and blood flow to coronaries)

Prognosis

- Ability to return to normal lifestyle
- Improvement in ventricular function
- Work capacity better than preoperative
- Risk of arrhythmias unchanged
- Minimal medications
- Complete and dramatic relief of chest pain.

Patency

Year	*Artery*	*Vein*
1	97	90
5	95	60
10	90	40

Recent advances

- Age limitation changed (CABG done in 80 years old also)
- Improved myocardial preservation
- Composite arterial grafts
- Sutureless anastomosis
- Endoscopic vein harvesting
- Intracoronary injections of vascular endothelial growth factor (VEGF)
- All major cities have the centres and expertise
- TMLR

TMLR (transmyocardial laser revascularisation)

- Severe CAD not treated by PTCA or CABG
- Emulates reptilian circulation in the mammalian heart
- No CPB
- CO_2 or excimer laser
- Limited thoracotomy

- Creation of channels 1 mm in size in the myocardium from epicardium to endocardium
- Allows perfusion of blood directly from the LV cavity to the intramyocardium
- Creation of intramyocardial channels, neoangiogenesis and denervation are the factors which improve perfusion to the ischaemic myocardium.

Composite arterial grafts

- Arterial grafts have long-term patency.
- Patients with total arterial grafts may not require future surgery.
- Arterial grafts are used in the shape of T or Y.
- It can revascularise almost all blocked major vessels.

Endoscopic vein harvesting

- Only two small incisions
- With help of a subcutaneous tunnel, the vein is dissected
- Requires expensive equipment
- Major branches are tied and smaller ones are controlled by pressure
- Cosmetic advantage
- Takes longer time

OFF PUMP CORONARY ARTERY BYPASS SURGERY

Procedure

- Beating heart surgery, no heart lung machine
- Sternotomy, stabilisation devices used
- Short-acting beta-blockers to reduce the heart rate.
- Proper positioning of the heart with mechanical stabilisation device for proper visual presentation
- CPB standby, all vessels can be bypassed by proper position of heart, quick recovery, early extubation
- No CPB, less trauma, reduce risk of bleeding and kidney failure, reduced hospital stay.

Advantages

- Avoids physiological stress associated with CPB.
- Aortic manipulation leading to neurological complications avoided. Whole body inflammatory response that occurs due to CPB is decreased.
- Incidence of postoperative renal failure due to CPB is decreased.

Future developments/Recent advances

1. Bypass graft coupling devices to facilitate anastomosis (clips, stents).
2. Normalising left ventricular geometry with intracardiac devices.
3. Myocardial protection during CPB (ventricular assist devices). Ischaemic preconditioning pharmacologically preoperatively.
4. **Vascular intimal hyperplasia:** Important cause of graft occlusion. Gene-based therapies to prevent this before grafting the veins.
5. Minimally invasive direct coronary artery bypass **(MIDCAB)** surgery: Anterior submammary incision. LIMA dissected down with the aid of a thoracoscope and grafted to the LAD.

ABDOMINAL AORTIC ANEURYSMS (AAA)

- Aneurysms are defined as a focal dilatation of at least 50% larger than the expected normal arterial diameter
- For AAA—transverse diameter 3 cm or greater

PEARLS OF WISDOM

Most common type of true aneurysm with high propensity to rupture and it is the 15th overall leading cause of death.

Risk factors for AAA (Key Box 44.21)

KEY BOX 44.21

MOST IMPORTANT RISK FACTORS OF AAA

1. Age: Peak incidence at 80 to 85 years
2. Male : Female ratio 4 : 1
3. Genetic: First-degree relatives: 11-fold increase in relative risk
4. Tobacco use: 8 : 1 preponderance of aneurysms in smokers
5. Other factors: 55% demonstrate *C. pneumoniae* by immunohistochemistry.

Site

95%—infrarenal aorta, 5%—suprarenal aorta.

Salient features

Fates of aneurysm

1. Infection
2. Rupture (free/contained)
3. Thrombosis
4. Embolism

Growth rates

< 5 cm diameter—0.3 cm/year > 5 cm diameter—0.5 cm/year.

Clinical features

Incidental finding

1. Pulsatile abdominal mass on palpation
2. Undergoing evaluation for another abdominal pathology
3. Surgery for unrelated abdominal or pelvic operation.

Classical triad

1. Sudden-onset midabdominal or flank pain
2. Shock
3. Presence of a pulsatile abdominal mass (expansile pulsation).

Findings on palpation

1. Firm mass
2. Expansile pulsation over the mass
3. If upper border is palpable, then the origin is probably infrarenal.

Diagnosis

1. **Radiograph**—Eggshell pattern of calcification
2. **Abdominal ultrasound**
   a. Detail of the vessel wall
   b. Presence of plaques
   c. Size
3. **CT—Most precise**
   a. Proximal and distal extent
   b. Amount and location of mural thrombus
   c. Calcifications
   d. Adjacent structures
4. **MRI/MRA**
   a. Less frequently used
   b. Used if patient has renal failure
5. **Contrast arteriography**
   Less frequently used

Complications (Key Box 44.22)

KEY BOX 44.22

COMPLICATIONS

1. Most frequent—nonfatal MI and renal failure
2. Bleeding
3. Most serious GI complication—ischaemia of the left colon and rectum
4. Lower extremity ischaemia
5. Ischaemic injury to the spinal cord or lumbosacral plexus
6. Postoperative sexual dysfunction
7. Deep vein thrombosis (DVT)

Management

Low-risk AAAs

- Followed with serial size measurements. Reduce expansion rate and rupture risk by conservative treatment such as:
  1. Smoking cessation
  2. Blood pressure control
  3. Reduction of cholesterol
  4. Risk factor modifications
  5. Drugs used: α-blockers, NSAIDs—inhibits elastase
- MMP inhibitors—doxycycline
- Repair is indicated when it is symptomatic or > 1 cm/yea growth.

High-risk patients

- Delay in repair until larger diameter
- Endovascular aneurysmal repair (EVAR) can be tried.

Techniques of open repair

1. **Transperitoneal approach**
2. **Retroperitoneal approach** is indicated in cases of hostil abdomen, suprarenal aneurysm, horseshoe kidney peritoneal dialysis, inflammatory aneurysm and in ascites
3. **Minimal incision aortic surgery**
   12 to 15 cm incision, 9 cm proximal to the umbilicus.

Endovascular aortic aneurysm repair

Stent-graft is introduced into the aneurysm through th femoral arteries and fixed in place to the nonaneurysmal aorti neck and iliac arteries with self-expanding or balloon expandable stents (Palmaz stents), or with barbs, pins o hooks.

Special considerations

1. Inflammatory aneurysm

- Incidence is about 5%
- Adherent to: Duodenum, IVC, left renal vein, ureters—the reason being lymphatic obstruction during aneurysm expansion and secondary fibrosis and infection from chronic contained rupture
- Rupture is uncommon (as it is often symptomatic and treated before rupture)
- Retroperitoneal approach is preferred.

2. Aortocaval fistula

a. Continuous abdominal bruit +
b. High-output cardiac failure
c. 'Steal' phenomenon—ischaemia to the lower limbs.

Treatment: Fistula closure followed by AAA repair.

3. Horseshoe kidney

a. Kidney usually fused anterior to the aorta
b. Left retroperitoneal approach
c. Endovascular repair is not possible.

RUPTURED ABDOMINAL AORTIC ANEURYSM

Types of rupture

Anteriorly—into peritoneal cavity
Posteriorly—into retroperitoneum

Clinical features

Back and abdominal pain, pallor, diaphoresis and syncope. If untreated, it is fatal in all cases.

Treatment

- Immediate surgical repair
- If patient is unstable + previously diagnosed or a pulsatile mass, he is transferred immediately to OT.
- If stable + questionable diagnosis, a CT and open surgical repair, control the haemorrhage + resuscitation + aneurysm repair. Early postoperative mortality rate is 45%.

Differential diagnosis of ruptured AAA

- Angina pectoris, perforated peptic ulcer
- Acute pancreatitis, acute cholecystitis
- Acute diverticulitis, mesenteric vascular occlusion
- Prolapsed lumbar intervertebral disc, sciatica

INTERESTING 'MOST COMMON' IN THIS CHAPTER

- The most common cause of respiratory insufficiency following chest trauma is pneumothorax.
- Most common cyanotic congenital heart disease is tetralogy of Fallot.
- Most common cause of cyanosis in newborn is transposition of great vessels.
- Most commonly performed open heart surgery all over the world is CABG.
- Most common type of true aneurysm with high propensity to rupture is abdominal aortic aneurysm.
- Most common type of AAA is infrarenal
- Most commonly done investigation for AAA is CT scan
- Most common noncardiac complication after repair of AAA is renal failure.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated.
- Mediastinal tumours, bronchogenic carcinoma have been included.
- Coronary artery bypass graft and abdominal aortic aneurysm have been discussed in more detail.

MULTIPLE CHOICE QUESTIONS

1. Fracture of the following rib is a marker of severe trauma:
A. First
B. Fourth
C. Eighth
D. Tenth

2. Treatment of posterior flail segment is:
A. Strapping
B. Open reduction and fixation
C. No treatment is required
D. External fixator application

3. Posterior flail segment does not require treatment because:
A. Scapula supports the flail segment
B. It does not cause complications
C. It heals by itself
D. It has no physiological implications

4. 'Internal pneumatic fixation' for flail chest is the term used for:
A. Insertion of a balloon into the chest
B. Internal fixation with screws
C. Endotracheal intubation and positive pressure ventilation
D. Valsalva manoeuvre

5. Tension pneumothorax should immediately be treated with:
A. Intercostal tube insertion
B. Needle thoracostomy
C. Thoracotomy
D. Thoracoscopic drainage

6. Features of tension pneumothorax include all of the following *except*:
A. Tachypnoea
B. Hypotension
C. Dull note on percussion
D. Tachycardia

7. Indications for thoracotomy in haemothorax include all of the following *except*:
A. Drainage more than 1000 ml
B. Drainage of more than 100 ml/hour for 4 hours
C. If clotted, haemothorax is suspected
D. Coagulation abnormalities

8. The intercostal drain can be removed in all of the following situations *except*:
A. Lung is fully expanded
B. Drainage < 100 ml
C. No air leak
D. The patient is on a ventilator

9. The following is diagnostic of surgical emphysema:
A. The lungs are emphysematous
B. Always follows surgery
C. Palpable crepitus
D. Infiltrates on chest X-ray

10. The following is true about mediastinal emphysema *except*:
A. The emphysema is mainly suprasternal
B. Mediastinal drain needs to be inserted
C. Pericardial crunching sounds can be heard on auscultation
D. Oesophageal and tracheal injury need to be ruled out

11. Nontraumatic rib fractures may be seen in all of the following *except*:
A. Hypoparathyroidism
B. Metastatic disease
C. Older patients after violent coughing
D. Osteogenesis imperfecta

12. The classic 'three' sign of coarctation of aorta is formed by all of the following *except*:
A. Dilated left subclavian artery
B. Narrowing of coarctation
C. Post-stenotic dilatation of aorta
D. Dilated pulmonary artery

13. Most common type of aneurysm with a propensity t rupture:
A. Abdominal aortic aneurysm
B. Carotid artery aneurysm
C. Radial artery aneurysm
D. Cerebral artery aneurysm

14. The most common cause of noncardiac complicatio after abdominal aortic aneurysm repair is:
A. Pulmonary insufficiency
B. Cerebral insufficiency
C. Renal insufficiency
D. Hepatic insufficiency

15. The most common cause of respiratory insufficienc following chest trauma is:
A. Pulmonary contusion
B. Pneumothorax
C. Flail chest
D. Haemothorax

ANSWERS

1 A	2 C	3 A	4 C	5 B	6 C	7 D	8 D	9 C	10 B
11 A	12 D	13 A	14 C	15 B					

45 Neurosurgery

- Classification of head injuries
- Primary lesions
- Secondary lesions
- Extradural/epidural haematoma
- Chronic subdural haematoma
- Raised intracranial pressure
- Fracture skull
- CSF rhinorrhoea
- Pott's puffy tumour
- Hydrocephalus
- Brain tumours
- Brainstem death
- What is new?/Recent advances

Introduction

Head injuries derive their importance because of the fact that many patients who die or who are disabled belong to the younger age groups. Head injuries account for 1% of all deaths, one-fourth of deaths due to trauma and they are responsible for **half of all deaths** from road traffic accidents. Majority of the patients are young, adult males.

HEAD INJURIES

Classification

I. Based on clinical type

1. Open
2. Closed

II. Based on type of injury

1. Blunt injury—acceleration, deceleration
2. Missile injuries

- The term **open head injury** is used to denote a type of injury in which there is a **fracture of the skull associated with tear of the dura and arachnoid**, resulting in cerebrospinal fluid leak either to the external environment or into one of the **potentially infective areas in the base of the skull, e.g. CSF rhinorrhoea or otorrhoea.**
- **A closed head injury** is one where there is **no such leakage.** The advantage of this classification is that it helps the treating physician to recognise a group of patients who are likely to develop an infective complication following the head injury and he can initiate measures to prevent it.
- **Blunt injuries,** depending on the severity of impact can result in an open or closed head injury. Missile injuries tend to result in an open head injury most often.
- The brain is protected by a bony box which has a **vault** and **base of the skull.** The **base** of skull in contrast to the vault is a **rough terrain** due to the various **bony prominences, ridges** and **foraminae**.
- This factor is important in causing extensive brain damage to the brain in acceleration/deceleration type of injuries. In addition to the linear acceleration/deceleration, **rotational acceleration** is also capable of producing damage to the brain as the **brain swirls about** inside the skull. Such injuries result in maximal damage at interfaces between structures of different densities such as **grey matter-white matter junctions.**

Pathology

The pathological changes due to trauma to the brain can be classified into primary and secondary.

I. Primary lesions

- Diffuse neuronal damage
- Shearing lesions
- Contusions and lacerations

II. Secondary lesions

- Swelling, haemorrhage
- Extradural haematoma

- Subdural, intracerebral haematoma
- Infection

I. Primary lesions (Key Box 45.1)

- **Diffuse neuronal damage is the most constant** feature of blunt injuries. Immediately after an injury no changes may be seen but changes begin **after 14 hours of injury and maximum effects** may last up to **one week. Prolonged unconsciousness** may follow injuries which produce only diffuse neuronal damage without any obvious macroscopic changes. Shearing lesions of the nerve fibres account for some severe injuries without any conspicuous changes to naked eye examination of the brain. A few important primary lesions are discussed below.
- **Cerebral concussion:** Alteration in consciousness without structural damage as a result of non-penetrating traumatic brain injury. There might be loss of consciousness, confusion and amnesia. These are the features. Widespread degeneration of white matter occurs without much changes in the nervous system cortex or brainstem. These patients have spasticity in all four limbs after injury and **when they regain consciousness**, they are found to be severely demented (Key Box 45.2).
- **Contusion** and **lacerations** are the obvious naked eye changes seen after injuries and were thought to be the main injuries before diffuse neuronal damage and shearing lesions were described. Contusions are seen on the summit of the gyri which get injured against the bone. **The overlying pia is torn and the blood seeps into the subarachnoid space.** A bleeding cortical vessel may result in the formation of the **acute subdural haematoma or intracerebral haemorrhage. Brain oedema** which develops surrounding the contusion and lacerations is the one that determines the outcome. Most often contusions are seen at the **tips of the frontal and temporal lobes, under surface of frontal and temporal lobes, over corpus callosum, superior and anterior surfaces of cerebellum and anterior surface of brainstem.**

KEY BOX 45.1

PRIMARY LESIONS

- Diffuse neuronal damage
- Cerebral contusion
- Cerebral laceration

KEY BOX 45.2

CEREBRAL CONCUSSION

- Temporary physiological paralysis of the nervous system
- Loss of consciousness
- Post-traumatic amnesia
- Recovery may be complete
- Some can develop complications

II. Secondary lesions (Key Box 45.3)

Brain swelling: This is a vague term applied to increase in brain bulk due to both oedema and venous congestion. It is aggravated by hypoxia or respiratory insufficiency which may be due to associated lung injury or obstruction to upper respiratory passages. Sometimes such a swelling can lead to severe brain compression which is difficult to relieve, since there is no single mass lesion.

PEARLS OF WISDOM

Malignant cerebral oedema has close to 100% mortality. This is more common in children.

Intracranial haemorrhage: Extradural or subdural haemorrhages may develop as a clean cut secondary event, even though bleeding may have started at the time of injury. These cause compression of brain, secondary rise in intracranial pressure and can cause death if not detected and treated early.

Infections: All open head injuries are liable to result in intracranial infection either as generalised meningitis or focal infection such as **subdural empyema or brain abscess, osteomyelitis of skull**. After closed head injuries, infection of a subpericranial blood clot may result in **Pott's puffy tumour.** When infection supervenes on an already injured brain, it may retard the recovery or may even lead to death. Hence, it becomes mandatory to treat all infections vigorously.

Cause of death in head injuries

- It is instructive to consider the pathological findings in fatal cases and to speculate the **deaths** which might have been prevented. For example, earlier many deaths which had occurred as a result of aggravation of brain swelling **due to hypoxia** could have been prevented by **ventilation** and **anti-oedema measures**. It should be emphasised that the role of the treating physician is to anticipate and take

KEY BOX 45.3

SECONDARY LESIONS

1. Brain swelling
   - Oedema
   - Venous congestion
   - Hypoxia
2. Intracranial haemorrhage
   - Extradural
   - Subdural
3. Infections
   - A. Open head injury
     - Generalised meningitis
     - Subdural empyema
   - B. Closed head injury
     - Pott's puffy tumour

appropriate measures to prevent the patient from succumbing to the secondary injuries (damages—seizures, hypoxia). **In extensive primary damage to the brain, apart from supportive treatment, one may have to wait and hope.**

- Extensive injury to vital areas such as diencephalon, or patients with **diffuse damage** are **not likely to survive**. These are patients who are unconscious from the time of injury with bilateral, dilated, fixed pupils, flaccidity in all 4 limbs and autonomic disturbances.
- Sometimes a head injury associated with extensive injuries to chest, abdomen or the limbs by their sheer severity can cause death.
- Intracranial complications such as haematomas, brain swelling, infection, and extracranial complications such as chest injury/metabolic abnormalities, if recognised and treated early, can go a long way in saving the life of the patients.

INTRACRANIAL HAEMATOMA

- Most of the head injuries are mild or minor and irrespective of how they are managed, the patient recovers on his own. All those who are unconscious, even if briefly, run the risk of respiratory obstruction. Some of the so-called trivially injured run the risk of developing an intracranial haematoma. Hence, all head injuries must be taken seriously. A **complicated head injury** is one where anyone of the **secondary pathological changes** may occur and threaten the life of the patient. **Uncomplicated** head injury is one where no **such events** occur. However, it could be a severe one where the unconsciousness is prolonged.
- These haematomas could develop in any one of the planes intracranially. Extradural (epidural), subdural, intracerebral haematoma, or a haemorrhagic contusion.
- The clinical presentation of these haematomas are due to either increase in the intracranial pressure or due to signs of cerebral compression. In the case of acute subdural haematoma or intracerebral haematoma, the clinical picture and the outcome of treatment is also dependent on associated brain damage.

EXTRADURAL/EPIDURAL HAEMATOMA (Fig. 45.1)

- The clot collects between the dura and the inner table of skull. A majority of them occur in the **middle cranial fossa,** since injury to **middle meningeal vessels (vein and artery) is the commonest cause.** However, about 20–25% of the extradural haematomas can occur in the frontal, parietal regions, at the vertex or in the posterior fossa. Injuries to the dural venous sinuses or a large diploic venous channel are the other causes for the formation of a haematoma. Depending upon the source of bleeding, the haematoma could collect rapidly (hyperacute type) or slowly over a period of a few hours to a few days and present as a **chronic lesion**. 60–80% of these patients have an associated fracture of the skull bone and only a few of them may present with classical symptoms with **lucid interval**. In the remaining patients, the initial picture can vary from an unconscious state to a fully conscious person with or without a history of post-traumatic amnesia. With the widespread availability and use of CT scan, the diagnosis has become much simpler nowadays. However, a few clinical features are worth mentioning.

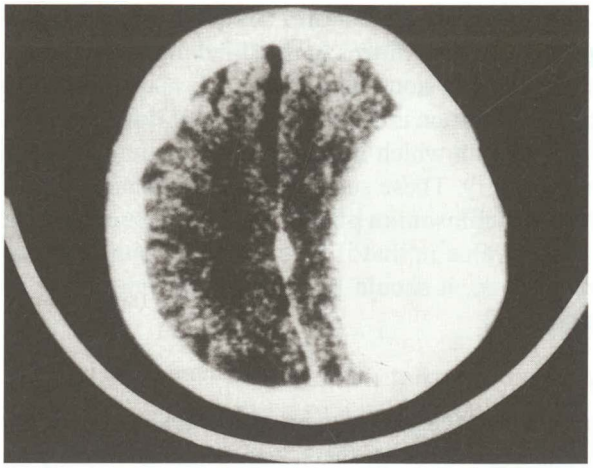


Fig. 45.1: CT scan showing extradural haematoma

1. Deteriorating consciousness level

- This is one of the hallmarks for the diagnosis of intracranial haematoma. The term **'lucid interval'** is used when a patient recovers from an initial period of unconscious state. Though in earlier days this was said to be associated with intracranial haematomas, it can occur in other conditions such as brain oedema, multiple contusions. To assess the consciousness level properly, instead of using vaguely defined terms such as semiconscious, obtunded, etc. the **'Glasgow Coma Score'** is widely used, to avoid observer errors in the observation of such patients.
- **Restlessness** in a previously quiet patient indicates **increasing intracranial pressure**, which again needs to be investigated. At the earliest appearance of focal neuronal deficit, the patient has to be taken up for exploratory burr holes.
- Progressive neurological deficit indicates cerebral compression and the manifestation may depend on the area of the brain affected.

2. Pupillary abnormalities

These are to be considered as a late manifestation. It is due to pressure by the **herniating temporal lobe on the ipsilateral third nerve at the tentorial hiatus**. In the **early stages** due to irritation of the nerve, there is **constriction.** Since it is a transient phenomenon, the early constriction goes unnoticed most of time. The patients are often detected in the **next stage**,

i.e. **pupillary dilatation,** caused by **paralysis of pupilloconstrictor fibres in the third nerve**. However, if the cerebral compression is unrelieved, this may go onto bilateral. Pupillary dilatation is due to **ischaemia of third nerve nucleus** at the midbrain which is caused by pressure on the posterior cerebral artery. These series of pupillary changes have been termed **Hutchinsonian pupils.** The dilated pupil has a definite localising value in that if an exploratory burr hole has been decided upon, it should be done on the side of the initially dilated pupil.

3. Autonomic disturbances

Bradycardia, though said to be a definite sign, is a late and not an early sign. Initially, there may be a rise in the pulse rate (tachycardia) which may progress to bradycardia, when the systolic blood pressure increases. At times there may be a rise in the diastolic pressure also. These **changes occur due to changes in the cerebral blood flow as a consequence of increased intracranial pressure**. Respirations become deep and slow rate (bradypnoea) and later patients may develop Cheyne-Stokes ventilation due to brainstem ischaemia.

PEARLS OF WISDOM

Cushing's triad of increased intracranial pressure (ICP).
- ***Bradycardia***
- ***Hypertension***
- ***Irregular respiration***

- Local scalp swelling is seen in more than half of the cases. Thus, examination of the head for any such swelling becomes important.
- Some of these patients may have a stiff neck either due to increased intracranial pressure or due to associated injury to neck muscles. Mild fever may, at times, occur and this sometimes confuses the observer. In such a case, the patients must be investigated with a definitive investigation like CT scan. If much time is not available, one should not hesitate to proceed to exploratory burr holes or a 'trauma craniotomy flap' has to be employed to rule out a haematoma.
- Though in adults, 'shock' is a rare complication of head injury, in children with intracranial haematoma and associated cephalhaematomas, due to volume depletion 'shock' may be encountered. Even in adults, if there is a large scalp injury which is not sutured immediately, shock can occur.
- Posterior fossa haematomas in any plane are dangerous because of the lesser space available for the haematomas. As a result of this, rapid brainstem compression can occur which may prove fatal. The availability of CT scan has made detection of these so-called 'unusual haematomas' more frequent. In a suspected case, even if **facilities are not available,** the treating physician should **explore the posterior fossa**, if the clinical features suggest haematoma, or if the skull X-rays show a fracture line extending across the occipital bone towards the foramen magnum.

Investigations (Key Box 45.4)

As has been pointed out earlier, the advent of CT scan of the head has made the diagnosis easier and more specific. However, it should be emphasised that **in the absence of CT scan,** if adequate clinical features point out to the **possibility of an intracranial haematoma**, the patient must be taken up immediately for an exploratory surgery rather than wait and allow him to develop irreversible brainstem damage. Since 60–80% of patients with an intracranial haematoma have a skull bone fracture, irrespective of his consciousness level has to be observed for at least 24–48 hours. Occasionally, one may have to resort to old investigations such as angiography not only to establish the haematoma but also to rule out associated dural venous sinus injury.

KEY BOX 45.4

INDICATIONS FOR SKULL RADIOLOGY
- Loss of consciousness
- Obvious depression on the skull
- Compound fracture
- Laceration or contusion of the scalp
- Focal neurological signs

TREATMENT OF HEAD INJURIES IN GENERAL

I. Resuscitation and support

1. Admission is indicated when:
a. Definite history of unconsciousness
b. Fracture temporal bone
c. Person who cannot be attended by the doctors immediately, i.e. no medical facilities nearby.
d. Post-traumatic seizures—patient should be admitted.

2. Casualty reception

a. Airway
- Mouth gag—to prevent tongue falling backwards.
- Endotracheal intubation with positive pressure ventilation. Hypoxia is an important cause of cerebral oedema which worsens the level of consciousness.

b. General assessment of patient
- To rule out abdominal injuries such as splenic rupture.
- Haemothorax—may need an intercostal tube.
- Long bone fractures

c. General assessment of the degree of shock by pulse, blood pressure monitoring and treatment.

d. Neurological assessment by Glasgow Coma Scale

1. Eyes open

Spontaneously	4
To speech	3

To pain	2
None	1
2. Best verbal response	
Oriented	5
Confused	4
Inappropriate words	3
Incomprehensible sounds	2
None	1
3. **Best motor response**	
Obeys commands	6
Localises the pain	5
Withdrawal to pain	4
Flexion to pain	3
Extension to pain	2 (severe damage with increase of ICP)
None	1

- Total score is 15; minimum score is 3. Any patient who has a coma score of 8 or less than 8 is said to be in coma.

II. Care of the unconscious

a. Ryle's tube aspiration or feeding
b. Care of the eyes—padding
c. Catheter for drainage of urine
d. Change of position to avoid bedsores.

III. Surgical treatment for extradural haematoma

Immediate surgery for removal of haematoma and relief of cerebral compression is a must. Extradural haematoma, in particular, is a neurosurgical emergency and patient survival will depend upon the speed with which the compression is relieved. It is not an exaggeration to state that even if decompression has to be done with unsterile instruments, at the bedside it may be worth the effort. In every neurosurgeon's career, at least one such situation might have occurred and a live patient may justify the means employed. Once consciousness is lost, pupils are dilated and decerebrate rigidity and periodic breathing develop, it may be only a few minutes that may be available to save the life of the patient and one should not wait and waste time. In the case of extradural haematoma the **outcome is dependent on the size of the haematoma and the stage in which** the patient was taken up for surgery. In the case of acute subdural and intracerebral haematoma it depends on associated brain damage. If the associated brain damage is very severe, patients succumbs to the brain damage (Key Box 45.5 and Fig. 45.2).

Acute subdural haematoma

Impact damage is more as compared to epidural haematoma. There is associated underlying brain injury. Symptoms are due to compression of underlying brain with midline shift, in addition to parenchymal brain injury.

Causes

- Parenchymal laceration bleed
- Torn cortical bridging vessel

KEY BOX 45.5

EXTRADURAL HAEMATOMA

- 3 cm vertical incision immediately above the midpoint of zygoma
- Strip the pericranium
- Burr hole with Hudson's brace
- Evacuate 'black-currant jelly' clot
- Extend the burr hole and control bleeding middle-meningeal artery by bipolar diathermy
- Dural hitch sutures to prevent stripping of dura

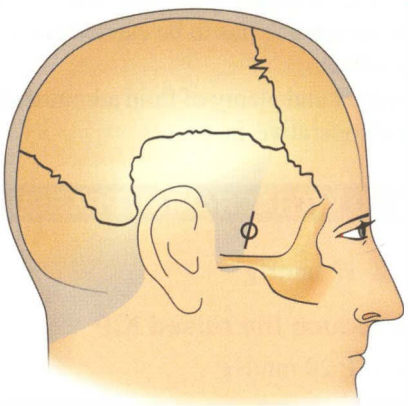


Fig. 45.2: Site of temporal burr hole

May occur in people who are on anticoagulant therapy.
Mortality—50–90%
Outcome is better if surgery is done within 4 hours.

CHRONIC SUBDURAL HAEMATOMA

- Common in old people
- In the elderly, the distance between the dura and the brain increases because of the shrinkage of the brain. Even a minor trauma can tear the cortical veins resulting in collection of blood
- Bleeding is never progressive and the blood in the subdural space slowly compresses the brain causing features of raised intracranial pressure (ICP).

Clinical features

- Elderly patients with history of minor trauma
- Bilateral headache, mental apathy
- Slowness, confusion—later alteration in the level of consciousness may progress to unconsciousness.
- Waxing and waning of the level of consciousness is seen in some patients. If such a history is elicited, one should always suspect chronic subdural haematoma.

Diagnosis

CT scan or, if feasible, MRI scan are the ideal investigations (cerebral angiography had been used and is still being used in

some centres where access to the latest imaging facilities are not available).

Treatment

- Burr hole and drainage of the haematoma usually under local anaesthesia or occasionally under general anaesthesia is often the practised mode of treatment.
- At times, the patient may need two or more burr holes to ensure adequate evacuation.
- If the brain fails to expand and obliterate the cavity, especially in older people or in persons with a very thick inner membrane, a large craniotomy and wide excision of the subdural membrane has to be carried out to remove the constricting effect.
- Adequate bedrest and plenty of fluid administration are also important postoperative measures.

RAISED INTRACRANIAL PRESSURE

Normal ICP is 8–12 mmHg

Measures to reduce the raised ICP

Aim is to keep ICP 20 mmHg

- Head and elevation
- Hyperventilation
- Sedation—with or without muscle relaxant
- Use of diuretics—furosemide, mannitol
- Thermoregulation
- Use of barbiturates—thiopentone—reduces brain metabolic rate
- Maintaining fluid and electrolyte balance
- Seizure control
- Steroids in severe head injury are associated with increased mortality and should not be used.

FRACTURE SKULL

Anterior fossa fracture

1. Fracture cribriform plate can result in CSF rhinorrhoea.
2. Fracture may extend to the orbit—subconjunctival haemorrhage.
3. Olfactory nerve involvement—partial anosmia.
4. Optic nerve may be contused or fracture may involve the optic foramen resulting in partial or total loss of vision.
5. Rarely, 3rd nerve palsy gives rise to dilated pupil.

Middle cranial fossa fracture

1. Epistaxis due to fracture venous/sphenoid sinuses
2. CSF from the ear: Blood mixes with CSF and so, does not clot.
3. 7th nerve palsy
4. Rarely 6th and 8th nerves are also involved.

Posterior cranial fossa fracture

1. Extravasation of blood in the suboccipital region causing boggy swelling in the nape of the neck.
2. 9th, 10th, and 11th cranial nerves may be involved.
3. **Battle sign:** Discolouration of skin and collection of blood occur in the region of mastoid process.

CSF RHINORRHOEA

- There should be a communication between the intradural cavity (subarachnoid space) and the nose.
- It indicates tear of the dura mainly in the basal region and a fracture involving paranasal sinuses—frontal, ethmoidal or sphenoidal.
- There is always an injury to a small portion of the brain. It (the portion of brain) plugs the tear, preventing the dura from healing. Thus, the rhinorrhoea persists for many days.
- This leads to complication, i.e. infection and meningitis.
- **Two types**
  1. **Traumatic:** It can be iatrogenic following surgery or can be post-traumatic (62 to 80%).
  2. **Nontraumatic:** It can be due to high pressure (hydrocephalus) or congenital.
- Confirmation of CSF rhinorrhoea is on the bed side by following clinical signs.
  1. **Ring sign:** Onto linen, ring of blood with 1 layer ring of clear fluid.
  2. **Reservoir sign:** Gush of CSF in certain position of the head.
- β_2 transferrin is the most accurate method.

Treatment

- Acetazolamide 250 mg three times a day
- Lumbar drainage can be done
- Prophylactic antibiotics
- If the rhinorrhoea persists, repair of the dural defect alone, (or) at times with a shunt procedure will be needed.

POTT'S PUFFY TUMOUR

- This is **subperiosteal infection** usually caused by osteomyelitis of the underlying skull.
- It is common in the frontal region and the **frontal bone is commonly involved.**
- The cause of infection is through frontal sinusitis.
- Another common cause of infection of a subpericranial haematoma following needle aspiration.
- It can also follow **chronic suppurative otitis media.**
- Pus collects in the subpericranial space and extradural plane, which communicate with each other (dumb-bell type abscess).
- It causes a **boggy swelling in the frontal region** and tenderness over the scalp.

- Pitting oedema over the scalp is conclusively called **Pott's puffy tumour**.
- Severe headache, vomiting and blurring of vision should clinch the diagnosis.

Treatment

1. CT scan to confirm the diagnosis
2. A burr hole and aspiration of pus can be done followed by 6–8 weeks of antibiotics.
3. In chronic cases, the wall of the abscess may have to be removed. The associated osteomyelitic skull bone requires a radical removal under cover of antibiotics.

HYDROCEPHALUS

Definition

This is a condition that occurs due to disturbances of CSF flow and imbalance between CSF production and absorption resulting in the accumulation of CSF and dilatation of ventricles.

CSF production

CSF is mainly produced by the choroid plexus of lateral ventricles by an active autoregulated process.

Daily production: 450 ml—total volume is 150 ml

CSF circulation (Fig. 45.3)

Lateral ventricle
↓ *via* foramen of Monroe
3rd ventricle
↓ *via* aqueduct of Sylvius
4th ventricle

CSF leaves the 4th ventricle by foramen of Luschka and Magendie to circulate over the convexity where it is finally absorbed over arachnoid granulations.

Aetiopathology

Hydrocephalus occurs due to **two reasons:**

- If there is overproduction of CSF, or CSF circulation
- If there is decreased absorption

A. Overproduction: True overproduction is rare and occurs in cases of **choroid plexus papilloma.**

B. Decreased absorption: Failure of CSF absorption is much more common due to infection and haemorrhage. Other causes being: Structural abnormality occurring in CSF pathway—tumour, congenital malformation such as aqueduct stenosis.

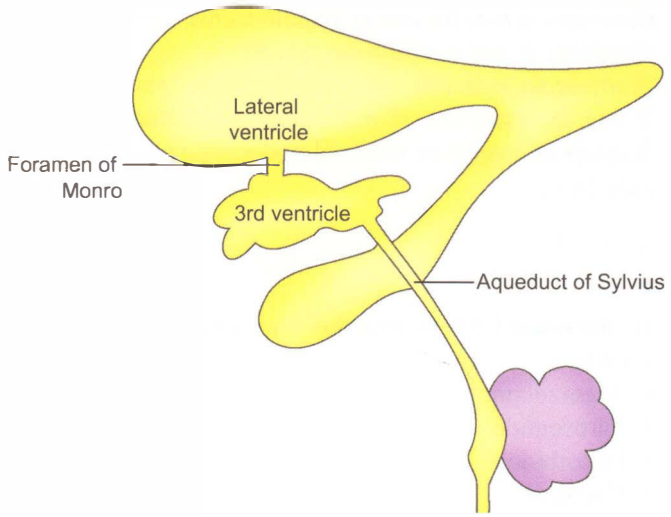


Fig. 45.3: Ventricular system

Types

1. **Communicating:** Due to obstruction in subarachnoid space.
2. **Noncommunicating:** Due to obstruction in ventricular system.

Clinical features

I. Infantile hydrocephalus

- Difficulty in delivery of large head (Key Box 45.6)
- Craniofacial disproportion
- Increase in head circumference more than 2 cm/month
- Scalp is thin, shiny and prominent veins
- Fontanelles: Bulging and tense especially on crying
- Sutures: Open, excessive irritability
- **Macewen's sign**—cracked pot sound on percussing over dilated ventricles.
- Inability to retain feeds, mental retardation, delayed milestones, hypothalamic disturbances.
- **Sun-set sign:** Weakness of upward gaze

II. Childhood/adult hydrocephalus

- By this time fontanelles have closed
- Features of increased intracranial pressure: Headache, nausea, vomiting.
- Irritability, indifference, apathy, drowsiness

KEY BOX 45.6

DIFFERENTIAL DIAGNOSIS OF LARGE HEAD

1. Megalencephaly	Intracranial pressure is normal
2. Chronic subdural haematoma	Enlargement of parietal region
3. Cerebral atrophy	May cause ventricular enlargement
4. Cerebral tumours	

- Blindness is not the sign of papilloedema
- Blindness is due to ophthalmoplegia
- Bradycardia, systemic hypertension, altered respiratory rate is due to distortion of brainstem. Untreated cases also develop unilateral or bilateral abducens palsy or upward gaze palsy.

Treatment

Aim

1. **To decrease CSF production** by using pharmacological agents:
   - Acetazolamide
   - Furosemide
   - Isosorbide
   - Glycerol
2. **Direct removal** of cause of obstruction
3. **Diversion** of CSF to another viscous for reabsorption by means of various shunt procedures.

SHUNTS

Ventriculoperitoneal shunt and ventriculoatrial shunt (Figs 45.4 and 45.5)

Complications of shunts (Key Box 45.7)

KEY BOX 45.7

COMPLICATIONS OF SHUNTS

1. Shunt obstruction
2. Shunt infection
3. Seizures
4. Extracerebral fluid collection
5. Subdural haematoma
6. Spontaneous pneumocephalus
7. Ascites

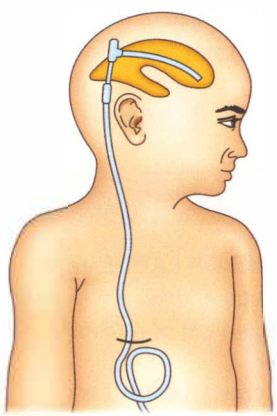


Fig. 45.4: Ventriculoperitoneal shunt

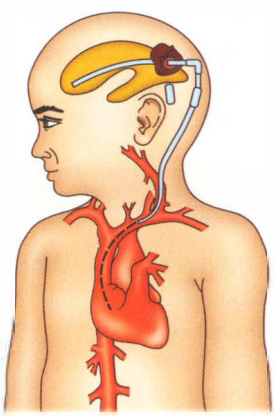


Fig. 45.5: Ventriculoatrial shunt

TUMOURS

Eight percent of all primary cancers arises in the CNS. In adults, it constitutes the sixth largest group of cancers and in children the CNS is the commonest site for solid tumours.

PEARLS OF WISDOM

Most adult brain tumours are supratentorial but 60% of tumours in children are infratentorial and arise in the posterior fossa.

Presenting feature

1. **Progressive neurological deficit:** In up to 2/3rds of the patients, a focal motor weakness is the most common presentation. This deficit arises from the direct neuronal damage or are due to compression on brain or cranial nerves by the tumour. Sudden change may be due to haemorrhage into a pre-existing tumour.
2. **Headache:** It occurs in over 50% of patients caused by raised intracranial pressure (ICP). Only 10% patients have the classic presentation of headache with early morning worsening, associated with nausea and a temporary relief by vomiting.
3. **Generalised or focal seizures** occur in 25%. Focal seizures may help to localise the site of tumour. A high index of suspicion is required for patients past the second decade with recent onset epilepsy.
4. **Other presentations** include visual disturbances, visual field defects, mental changes, etc. depending on the site of tumour.

Investigations

1. **CT scan:** The best initial investigation. This shows where the tumour alters the attenuation of the X-ray beam as it passes through the brain. It also shows distortion of the ventricular system or obliteration of the pattern of the sulci.
2. **MRI:** Often gives extra information and may eventually supersede CT completely. It is the investigation of choice for imaging the posterior fossa and the spine as it can 'see through' bone.
3. **X-rays** are only of anecdotal interest picking up incidental calcifications or erosion of parts of the skull. Newer imaging modalities including SPECT and PET scans are increasingly being used in higher centres.

SPECIFIC TYPES OF BRAIN TUMOURS

Gliomas

- Astrocytoma
- Oligodendroglioma
- Mixed tumours

Astrocytoma: WHO classification

Grade I : Pilocytic
Grade II : Diffuse
Grade III : Anaplastic
Grade IV : Glioblastoma multiforme

- This is the **most common primary brain tumour** arising from the supporting glial cells and diffusely infiltrates brain tissue early on.
- **Two grading systems** are available: WHO and St Anne/ Mayo. Grade 1 and Grade 2 (low grade) tumours are slow growing and compatible with good quality survival. Grades 3 and 4 are rapidly growing. Grade 4 tumours are also called glioblastoma multiforme are highly radio and chemo-resistant with median survival of only 12 months even with optimal treatment. Even low grade astrocytomas may evolve over time into secondary glioblastomas.

Glioblastoma Multiforme

- Most common in 5th and 6th decade.
- Treatment includes surgery to confirm the diagnosis and achieve a macroscopic excision, followed by a high dose (60 gy) of irradiation.
- Chemotherapy wafers impregnated with carmustine must be inserted.

Oligodendroglioma

- This type of glioma is usually a **slow-growing tumour** and over half arise in the frontal lobes.
- There may be a **history of epilepsy** or even focal neurological signs of many years' duration.
- They may show **calcification,** both microscopically and macroscopically.

Ependymoma

- These glial tumours arise from cells that line the ventricles of the brain and the central canal of the spinal cord.
- They are **most common in the fourth ventricle** in children and young adults where they block CSF flow and often present with hydrocephalus.

Embryonal tumours

- Primitive neuroectodermal tumours (PNETs) are a group of **highly malignant tumours** of which the cerebellar PNET or medulloblastoma is the archetype.
- Commonest in children and young adults. It may originate from primitive cell nests which have undergone malignant transformation. Medulloblastoma is the most common brain tumour in children.
- The patient presents with truncal ataxia, headache, vomiting and sometimes diplopia. All PNETs are prone to spread within the CNS producing '**sugarcoating metastasis**' which are best seen on MRI of the spine.

Schwannoma (neurilemmoma)

- Peripheral neurons get their myelin sheaths from Schwann cells. Occasionally, these form slow growing benign tumours on cranial or spinal nerves.
- The cranial nerve most affected is the **vestibular division of the 8th cranial nerve** and the tumour is then usually called **acoustic neuroma**.

PEARLS OF WISDOM

Even though it does not arise from the acoustic division and is not a neuroma, it is still called an acoustic neuroma.

- It causes progressive deafness, hydrocephalus and ataxia It is slow growing. Less often it may involve the trigeminal or the vagus nerve.

Meningioma

- Eighty percent are supratentorial
- These tumours arise from the arachnoid layer of the meninges and the arachnoid villi. They are commonest over the falx and the convexity of the skull but rarely may also arise from the skull base (especially from the sphenoid wing and olfactory groove) or inside the lateral ventricle.
- Commonest in middle-aged women and may occur at the site of a previous radiation field. Most are benign and slowly growing. Signs occur based on the site of tumour (Key Box 45.8).
- They tend to provoke endosteal hypertrophy or exostosis of the overlying skull and is still occasionally detected on an incidental skull film or even by palpation of the skull. Patient with neurofibromatosis type 2 often have multiple meningiomas. Rarely, they are malignant.

Pituitary tumours

- 10–15% of all intracranial tumours
- Majority are benign adenoma
- Prolactinoma—30%
- Nonfunctioning adenoma—20%
- GH secreting adenoma—15%
- ACTH secreting adenoma—10%

KEY BOX 45.8

SITES OF MENINGIOMA

• Convexity of skull	:	20%
• Parafalcine arc	:	20%
• Sphenoid ridge	:	10%
• Tuberculum sella	:	10%
• Olfactory groove	:	10%
• Ventricle	:	2–5%

- May produce **mass effect**—bitemporal hemianopia or cranial nerve dysfunction.
- **Endocrine dysfunction** such as galactorrhoea, primary/secondary amenorrhoea, Cushing's syndrome, acromegaly.
- **Pituitary apoplexy**—results in sudden onset of headache, visual loss, ophthalmoplegia and altered conscious level. Caused by haemorrhagic infarction of a pituitary tumour.

Other tumours

They are pineal region tumours, pituitary adenomas, craniopharyngiomas, choroid plexus tumours, etc.

Metastases

One-fourth of all cancer patients have intracerebral metastases at the time of death. Common primary sites include the bronchus (50%), breast (15%), and melanoma (10%).

Treatment

They can be divided into medical and surgical line of treatment.

I. Medical

- Acutely raised intracranial pressure (ICP) is treated with **IV mannitol 0.5–1 g/kg body weight**.
- Hydrocephalus can be relieved using a **CSF diversion** system (closed external ventricular drainage or a **ventriculoperitoneal shunt**).
- Seizures are treated with **lorazepam and phenytoin** commonly and maintained on phenytoin.
- **Corticosteroids**, especially dexamethasone 4 mg qid is given to reduce symptoms of raised ICP, which may make surgery easier.

II. Surgery

- This is the **mainstay of treatment**. The aim of surgery is to obtain a complete tumour excision without producing a neurological deficit.
- This is not always possible due to the site of the tumour, in which case compromises must be made and a **debulking procedure** is done.
- Any residual tissue may be observed or treated with adjuvant radiotherapy.
- **Stereotactic guided surgery** is now a well-established concept in brain surgery allowing more targeted treatment of the lesion with minimal surrounding damage.

III. Radiotherapy

- Intracranial tumours are relatively **radioresistant**, and radiotherapy is primarily a palliative treatment.

IV. Chemotherapy

- No definite benefit of chemotherapy for the treatment of brain tumours. **Temozolomide** is a promising new drug for the treatment of brain tumours.

Outcome

Surgery offers good prospects for the treatment of benign brain tumours such as meningioma or pituitary adenoma, but outcome of treatment of malignant tumours is still poor.

TRIGEMINAL NEURALGIA

- Superior cerebellar artery or multiple sclerosis plaque can cause this condition.
- Severe episodic lancinating facial pain occurring in the distribution of V cranial nerve.
- Vascular compression of the nerve near the root entry zone, multiple sclerosis may be causative factors.
- Investigation—MRI

Treatment

- Carbamazepine or gabapentin
- 75% will not respond to medical treatment.
- Surgery: Refractory cases
  1. Percutaneous glycerol injection, radiofrequency ablation, thermocoagulation, balloon compression, local nerve block.
  2. Microvascular decompression *via* craniotomy
  3. Stereotactic radiosurgery

BRAINSTEM DEATH

Irreversible loss of consciousness, loss of brainstem reflexes and apnoea.

Diagnosis of brainstem death is done in three stages:

1. Identification of the cause of irreversible coma.
2. Exclusion of reversible causes of coma.
3. Clinical demonstration of absence of brainstem reflexes.

Brainstem reflexes

- Pupillary reaction to light, corneal reflexes, vestibular ocular reflex, cough reflex, gag reflex, motor response to pain.
- Apnoea test—apnoea despite a CO_2 increase to > 6.65 kPa or 50 mmHg.
- All reflexes must be absent and are tested independently twice by 2 doctors.

A FEW CT SCANS OF HEAD INJURIES (Fig. 45.6 to 46.14)

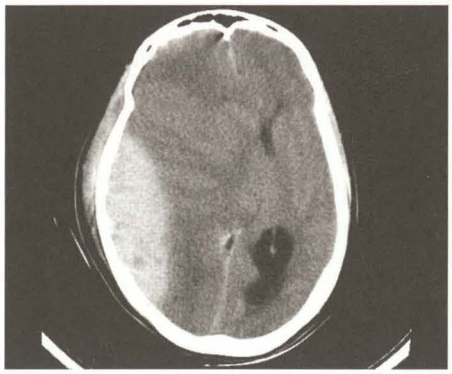


Fig. 45.6: Right temporo-parietal extradural haematoma(EDH) showing mass effect with midline shift with subfalcine herniation

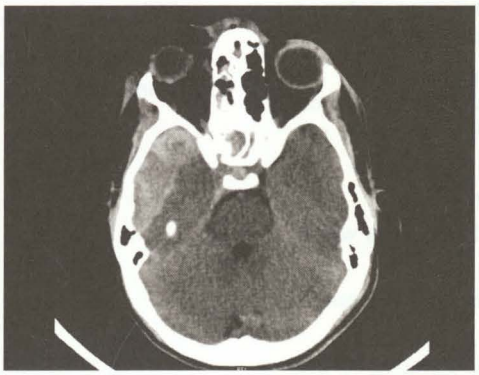


Fig. 45.7: Right temporal extradural haematoma

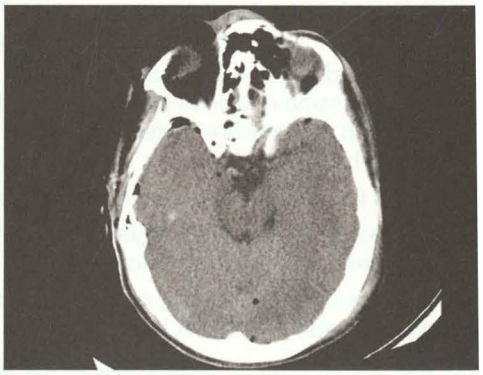


Fig. 45.8: Postoperative scan-no EDH. Post right temporal craniotomy with pneumocephalous

Fig. 45.9: Left frontoparietal acute subdural haematoma. Left frontal and temporal nonhaemorrhagic contusion with subfalcine herniation with midline shift

Fig. 45.10: Traumatic right more than left frontoparietal and anterior interhemispheric subarachnoid haemorrhage

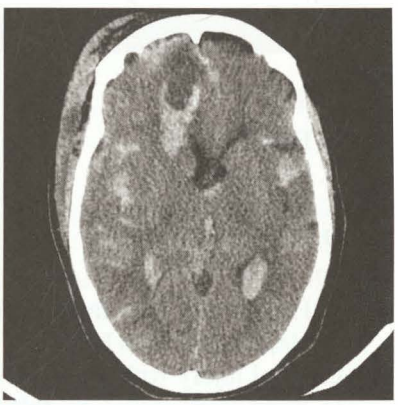


Fig. 45.11: Diffuse axonal injury—right frontal, left temporal contusion, right frontoparietal region—subarachnoid haemorrhage and intraventricular haemorrhage

Contributed by Dr. Sunil Upadhyay — Asst. Prof., Dept. of Neuro Surgery, KMC, Manipal

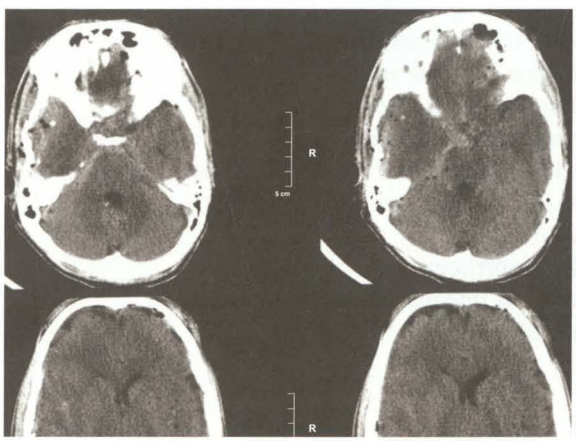


Fig. 45.12: Right temporal depressed fracture with temporal EDH with pneumocephalous

Fig. 45.13: Left frontoparietal hypodence area suggestive of left middle cerebral artery stem infarct

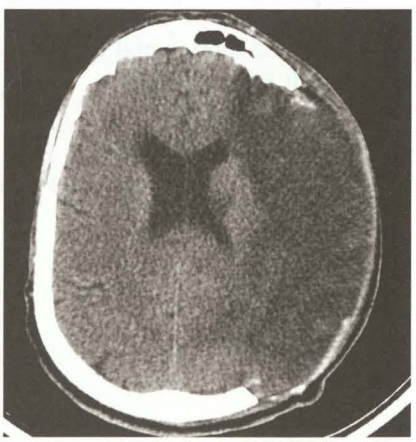


Fig. 45.14: Left FTP decompressive craniectomy brain herniation outside the skull

VERY IMPORTANT WISDOM LINES IN HEAD INJURY

- If an exploratory burr hole has been decided upon, it should be done on the side of the initially dilated pupil.
- Bradycardia is due to raised intracranial pressure.
- If hypotension responds to volume replacement in trauma centre in a comatose patient, the coma is most likely not due to head injury.
- Admit the patient if there is fracture temporal bone.
- Hypoxia is an important cause of cerebral oedema which worsens the level of consciousness.
- Steroids in severe head injury are associated with increased mortality and should not be used.
- CSF rhinorrhoea indicates tear of the dura mainly in the basal region and a fracture involving paranasal sinuses.
- Glasgow Coma Score of less than 8 means patient is in coma.

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated
- Trigeminal neuralgia, brainstem death have been included.

MULTIPLE CHOICE QUESTIONS

1. Following is not the feature of Glasgow Coma Scale:
A. Eyes opening
B. Motor response
C. Verbal response
D. Sensory response

2. Closed head injury include following *except:*
A. Head injury with fracture skull
B. Head injury with black eye
C. Head injury with facial nerve palsy
D. Head injury with CSF rhinorrhoea

3. Which are the following is included under secondary lesions following head injury?
A. Diffuse neuronal damage
B. Contusions
C. Lacerations
D. Swelling

4. Post-traumatic amnesia is a feature of:
A. Raised intracranial tension
B. Fall from a height
C. Fracture skull
D. Cerebral concussion

5. Which is an important cause of brain swelling following head injury?
A. Infection
B. Oedema
C. Acidosis
D. Bleeding

6. The cause of extradural haematoma is bleeding from:
A. Venous sinuses
B. Cavernous sinus bleeding
C. Basal veins bleeding
D. Middle meningeal vessels bleeding

7. Lucid interval is typically seen in:
A. Extradural haematoma
B. Acute subdural haematoma
C. Chronic subdural haematoma
D. Pontine haematoma

8. Which nerve is paralysed after herniation of temporal lobe in extradural haematoma?
A. Occulomotor nerve
B. Ophthalmic nerve
C. Trigeminal nerve
D. Facial nerve

9. Pupillary dilatation following head injury is due to ischaemia of the third nerve caused by:
A. Middle cerebral artery
B. Posterior cerebral artery
C. Inferior cerebral artery
D. Anterior cerebral artery

10. Following are definite indications for admission in a head injury patient *except:*
A. Fracture skull
B. CSF rhinorrhoea
C. History of unconsciousness
D. Scalp bleeding

ANSWERS

1 D	2 D	3 D	4 D	5 B	6 D	7 A	8 B	9 B	10 D

46 Principles of Radiology

- Barium swallow
- Barium meal
- Barium meal follow through
- Barium enema
- Enteroclysis
- Angiography
- Splenoportography
- Sialography
- Computed tomography
- Ultrasonography
- Magnetic resonance imaging
- Peripheral venography
- Myelography
- Interventional radiology
- PET scan
- Virtual colonoscopy
- What is new?/Recent advances

Introduction

These radiological investigations have become an integral part of the management of the patients. A clear understanding of these will help the students to read the images accurately, interpret well and score good marks.

BARIUM STUDIES

This is the study of the gastrointestinal tract by instillation/ingestion of barium suspension made up of/made from pure **barium sulphate**.

BARIUM SWALLOW (Fig. 46.1)

It is the contrast study from the oral cavity up to the fundus of the stomach.

Indications

- Dysphagia and obstruction
- Odynophagia
- Assessment of mediastinal masses
- Motility disorders of oesophagus—achalasia, scleroderma.

Relative contraindications

- Tracheo-oesophageal fistula
- Perforation

Procedure

- One mouthful of contrast media is given and the act of deglutition is observed fluoroscopically. After a mouthful of barium, films are exposed to the region of interest.

Fig. 46.1: Barium swallow: Opacified thoracic oesophagus showing an intraluminal mass with mucosal irregularity in its lower third suggestive of malignancy

Interpretation of study

1. Malignant obstructions are seen as annular constrictions, shouldering cranial and caudal to the lesion, mucosal destruction, ulceration and fistulae formation.
2. Benign strictures are long segment narrowings with no mucosal abnormalities.

3. Achalasia cardia is evident as 'rat tail' appearance of the lower end of the oesophagus, with gross dilatation of the oesophagus proximally and thin streaks of contrast entering the stomach.
4. Scleroderma shows dilatation, atonicity, poor or absent peristalsis and free gastro-oesophageal reflux.

BARIUM MEAL (Fig. 46.2)

This is the radiological study of oesophagus, stomach, duodenum and proximal jejunum.

Indications

- Symptoms of vomiting, epigastric pain, heart burn, dyspepsia
- Upper abdominal mass
- Gastrointestinal haemorrhage
- Gastric or duodenal obstruction
- Malignancies

Contraindications

- Suspected perforation
- Suspicion of aspiration
- Large bowel obstruction

Procedure

An undiluted barium suspension is given and deglutition is seen under fluoroscopy. Once barium reaches the stomach, the patient is rotated so as to coat the entire stomach and filming is done. More barium is given to distend the stomach wall. Filming is done as contrast enters the duodenum and opacifies proximal jejunum.

Interpretation of study

1. **Hiatus hernia** is evident as presence of the stomach above the oesophageal hiatus. In addition gastro-oesophageal reflux will be evident. Mucosal ulceration and strictures may be demonstrable in long-standing cases.
2. **Gastric and duodenal ulcers** appear as projections from the normal contour with pooling of contrast. Benign **ulcers** usually **project out** and have the mucosal folds radiating up to the edge of the ulcer. Deformity of the stomach and duodenal cap are seen in chronic stages.
3. **Bezoars of stomach** are seen as radiolucent masses in the stomach and the barium fills the crevices between the particles forming a characteristic appearance.
4. **Infantile hypertrophic pyloric stenosis:** Thin **streak** of barium is seen extending across pylorus—indentation of barium-filled antrum is seen.

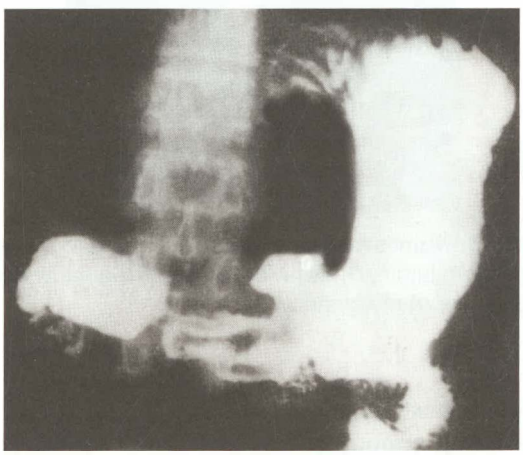


Fig. 46.2: Barium meal: Contrast opacified stomach demonstrating a projection from lesser curvature due to a benign gastric ulcer (niche)

BARIUM MEAL FOLLOW THROUGH

It is the radiographic examination of the GIT—oesophagus, stomach, small bowel and ileocaecal junction, by oral administration of contrast media.

Indications

- Symptoms of small bowel disease such as diarrhoea, abdominal pain, weight loss, Crohn's disease.
- Small bowel obstruction (chronic)
- Gastrointestinal bleeding
- Palpable mass possibly involving the small bowel
- Malabsorption

Contraindications

- Colonic obstruction
- Suspected perforation

Procedure

A small mouthful of barium is given after which the stomach is studied and films are taken. 500–800 ml barium is then given to the patient and filming is done after 15–20 minutes to demonstrate the jejunum and the proximal ileum. Subsequently, films of the ileum and ileocaecal junction are taken.

Interpretation of study

1. Intestinal malignancies such as lymphomas are evident as strictures which may be short or long segment with or without proximal dilatation. The mucosa shows irregularity and ulcerations. Fistulous communications may be evident. Displacement of bowel loops in large extraluminal masses may be seen.
2. Inflammatory strictures are evident as sites of narrowing with mucosal abnormality and ulcerations, proximal dilatation and mucosal fold thickening.

Complications

- Perforation of the bowel
- Aspiration

BARIUM ENEMA

This is the radiographic study of the large bowel by administration of contrast media through the rectum.

Types

Single contrast barium enema and double contrast barium enema.

Indications

1. Change in bowel habit
2. Melaena
3. Mass suspected to be arising from colon
4. Features of large gut obstruction (subacute)

Contraindications

1. Toxic megacolon
2. Pseudomembranous colitis
3. Rectal biopsy done recently (procedure withheld for 7 days)

Procedure

- Bowel is prepared with low residue diet, purgation and cleansing water enema. High density barium suspension is allowed to flow up to the ileocaecal junction and reflux into the terminal ileum. Single contrast filming is done. The patient is asked to evacuate the barium and a post evacuation film is taken. Once barium is evacuated properly, air insufflation is carried out so as to distend colon up to the ileocaecal junction.
- Filming is done to demonstrate the double contrast of large bowel with additional spots of hepatic, splenic flexures and rectosigmoid junction in oblique positions so as to open up these regions.

Interpretation of study

A few examples are:

1. Ulcerative colitis
- Loss of haustral pattern
- Fine granularity of mucosa
- Fine ulcerations resulting in spiking of colonic surface
- Strictures
- Pipe stem colon, increase in presacral space

2. Malignant lesions
- Circumferential/eccentric growth narrowing the lumen
- Hold up of barium proximal to the lesion
- Mucosal abnormality—ulcerations

3. Tuberculosis

Ileocaecal region is the commonest site. Deformed, elevated caecum, stricture and ulceration involving ascending colon and ileum.

4. Crohn's disease

Multiple ulcerations, thickening and distortion of valvulae conniventes, short or long strictures, cobblestone pattern and separation of bowel loops are the features.

5. Malabsorption

Dilution of barium, segmentation of the column of barium 'Moulage sign' (barium in a featureless tube) and jejuna dilatation are the findings.

SMALL BOWEL ENEMA OR ENTEROCLYSIS

(Fig. 46.3) (Key Boxes 46.1 and 46.2)

This is the radiological study of the small bowel (from jejunum to the ileocaecal junction) by intubation of the jejunum and instillation of contrast media through the tube.

Procedure

- Bilbao Dotter tube is inserted with the guide wire through one of the nostrils and advanced caudally with the swallowing action till the tip reaches the stomach. The tube is then advanced through the antrum of the stomach to the pyloric canal. Then it is advanced under fluoroscopic guidance to about 4–5 cm distal to the Trietz ligament (duodenojejunal junction).

Fig. 46.3: Small bowel enema showing stricture terminal ileum

KEY BOX 46.1

INDICATIONS
- Mechanical obstruction
- GIT bleeding
- Tumours of small intestine
- Unexplained abdominal pain
- Diarrhoea

KEY BOX 46.2

CONTRAINDICATIONS

- Complete obstruction
- Suspected perforation
- Massive dilatation of small bowel
- Duodenal obstruction
- Gastrojejunostomy

- 200 ml barium suspension is injected at a rate of 75 ml/min followed by 5% of methylcellulose at a rate of 100 ml/min. The head of the barium column is followed with intermittent fluoroscopy and films exposed wherever necessary.
- Ileocaecal spot films are taken when the junction is opacified and distended.

Interpretation of study

1. Normal small bowel shows a decrease in calibre from jejunum to ileum and the change of prominent valvulae conniventes to featureless ileum is evident.
2. Malignancies and lymphomas show evidence of strictures, proximal dilatations and mucosal abnormality. Large mesenteric nodal masses displace the bowel loops.
3. Strictures and ulceration of terminal ileum: Dilatation of the segment proximal to the narrowed segment and conical shrunken caecum are seen in ileocaecal tuberculosis. In later stages, ileal strictures, fistulae, etc. may be seen.

Complications

- Perforation
- Inspissation of barium
- Transient bacteraemia

ANGIOGRAPHY (Figs 46.4 and 46.5)

Definition

This is the study of blood vessels by injection of a contrast medium containing iodine into the vessel.

Indications

1. Primary vascular diseases such as vaso-occlusive disease, aneurysm, arteriovenous malformation (AVM).
2. Vascularity assessment of a tumour.
3. Congenital vascular conditions such as coarctation.
4. Percutaneous interventional vascular procedures.

Contraindications

1. Bleeding tendencies
2. Skin infections at site of entry
3. Cardiovascular disease such as recent myocardial infarction, overt congestive cardiac failure.
4. Hepatic failure

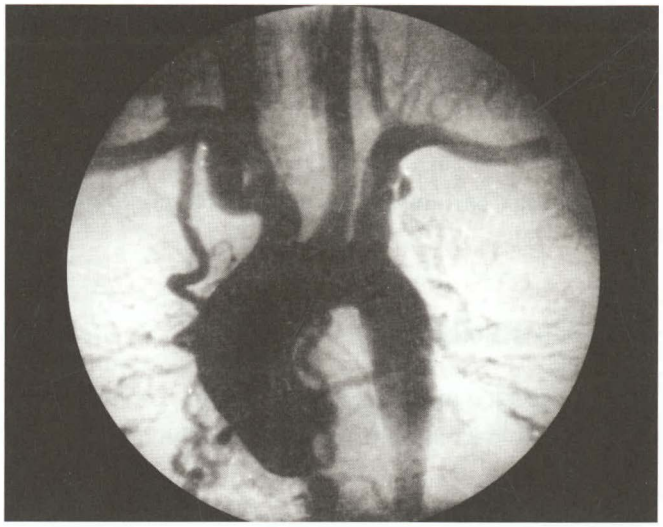


Fig. 46.4: Arch aortogram: Contrast in the arch of aorta demonstrating the major vessels arising from it

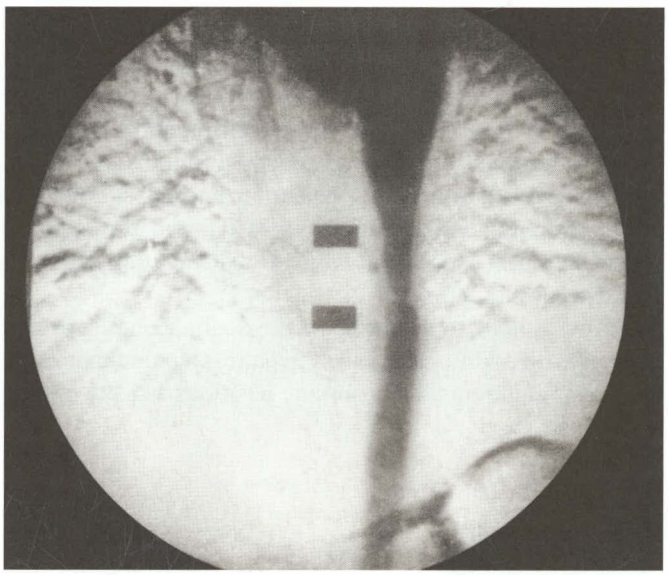


Fig. 46.5: Aortogram: Contrast in the thoracic aorta showing narrowing

Procedure

- Local anaesthesia at site of puncture is preferred except in children or restless patients, wherein general anaesthesia is preferred. Using a Seldinger needle the artery is punctured.
- The catheter of appropriate dimension is placed into the artery and negotiated into the desired vessel to be studied. Contrast is injected and filming is done.

Puncture sites

Femoral artery, axillary artery and brachial artery.

Interpretation of study

1. Aneurysms are seen as focal dilatations of vessel or projecting from the main vessel through a neck.

2. Tumour vessels show abnormal branching pattern, vascular encasement, displacement, arteriovenous shunting and pooling of contrast in the lesion.
3. AVM shows evidence of a dilated feeding artery/abnormal blush and early draining vein.
4. Vascular occlusions are seen as abrupt or gradual tapering of vessel with collateral supply distally.

Complications

- Damage to arterial walls at the site of puncture
- Severe hypotensive reactions
- Thrombosis of arteries, catheter clot embolus, haematoma at puncture site
- Vagal inhibition
- Allergic reactions to contrast
- Damage to nerves and to organs

SPLENOPORTOGRAPHY (Fig. 46.6)

This is the contrast study of the portal venous system by percutaneous splenic puncture.

Indications

- Demonstration of the anatomy of the portal system in patients with portal hypertension prior to surgery
- Check the patency of a portosystemic anastomosis

Contraindications

- Abnormal prothrombin time
- Ascites

Procedure

Splenoportography needle is introduced into the spleen in the midaxillary line under local anaesthesia. After confirming the placement of needle in the splenic pulp, injection of iodine containing contrast media is done and films are taken to includ the splenic vein, portal vein and the portal radicles in liver.

Fig. 46.6: Splenoportography showing dilated splenic vein

Interpretation of study

- Contrast injected into the splenic pulp drains into the hilun through the splenic radicles and then through the splenic vein and portal vein into liver.
- Thromboses are seen as filling defects in the contras opacified splenic and portal vein.
- In severe portal hypertension multiple collaterals are seer as branching channels from the normal pathway.

Complications

1. Haemorrhage
2. Perforation of adjacent structures (pleura, colon)
3. Splenic rupture
4. Infection

SIALOGRAPHY

This is the contrast opacification of the duct and the glandular acini by cannulating the ducts of salivary glands.

Indications

Pain, recurrent swelling

Contraindications

Acute infection or inflammation

Procedure

- The orifice of the parotid duct or submandibular duct is cannulated depending on the indication. Iodine-containing contrast medium is injected.
- Injection is terminated **immediately, if any pain is experienced**.
- Films are taken to demonstrate the duct and the glandular branching pattern.
- Post-secretory films are taken 5 minutes after administration of a sialogogue to demonstrate sialectasis.

Interpretation of study

1. Stones in the gland are identified as filling defects and those in the ductal system result in proximal obstruction and dilatation.
2. Glandular enlargement due to inflammation result in pooling of the contrast injected.
3. Tumours of the salivary gland result in irregular filling defect with duct distortion and pooling of contrast medium.

Complications

- Pain
- Damage to duct orifice
- Rupture of ducts
- Infection

COMPUTED TOMOGRAPHY (CT)

This is an imaging procedure where detailed information is obtained from thin sections in collimated X-rays.

Indications

- Structural evaluation of intracranial lesions (Fig. 46.7)
- Detailed evaluation of lung, mediastinal pathologies
- Intra-abdominal and pelvic masses where exact site of origin and relation to adjacent structures can be evaluated.
- Extra-osseous and soft tissue extension of bone tumours.
- Vascularity of the normal organ and the abnormal tissue can be evaluated and compared.

Contraindications (relative)

- Pregnancy
- Restless patients

Fig. 46.7: Cranial CT: Hypodensity of the left (L) frontoparietal cerebral parenchyma suggestive of a (L) middle cerebral artery territory infarct

Interpretation of images

- Structures imaged appear densely white to densely black depending on the absorption of X-rays and the emerging resultant X-rays which are detected. **The composite picture is actually a collection of Hounsfield numbers.** Each Hounsfield number being assigned a specific shade of grey, thus producing a picture that might be easily understood. Some of the common densities to be encountered in practice are as follows (Hounsfield units = HU):

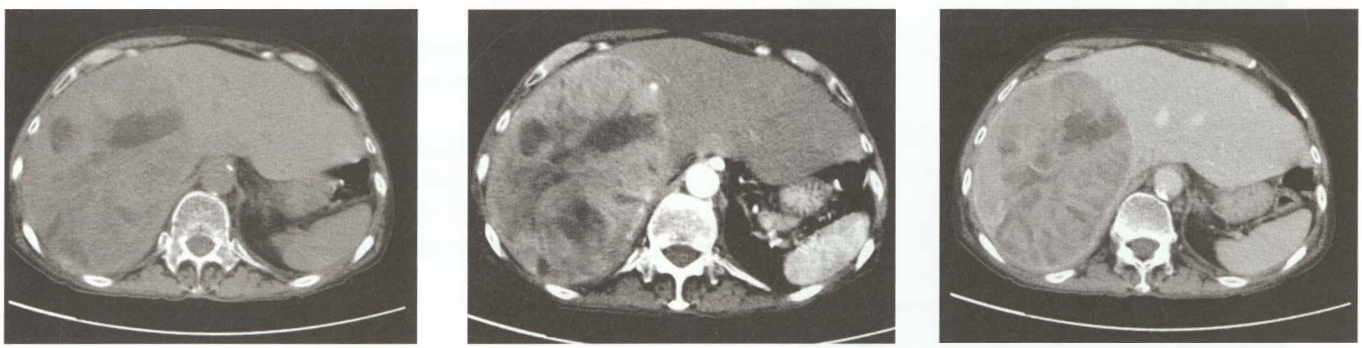
Air	–1000 HU
Fat	–50 to –100 HU
Water	0 HU
CSF	0 to +3 HU
White matter	+22 to +32 HU
Grey matter	+36 to +46 HU
Clotted blood	+60 to +80 HU
Calcification bone	+80 to +1000 HU

- In order to increase the contrast that may exist between the structures in the body, **intravenous contrast (iodine containing) is administered.** Certain tissues show enhancement of their density and various pathologies also show fairly characteristic contrast uptake patterns (Figs 46.8 to 46.11).
- In abdominal scanning, **oral contrast** is administered to the patient before the procedure, to enable the operator to accurately separate the bowel loops from the other intra-abdominal structures.
- The advantages of CT over conventional radiology are that it can visualise extremely small pathology, not evident on conventional films, is cost-effective as multiple X-ray films and procedures can be avoided. It is noninvasive and the radiation levels applied to the patient are extremely low.

ULTRASONOGRAPHY (Figs 46.12 and 46.13)

Principle

This imaging modality is based on the **piezoelectric effect** which is the property of certain substances to convert **electrical**



Figs 46.8 to 46.10: CT scan axial section through liver shows large mass in plain scan. Brightly enhancing in arterial phase of contrast study. Finally wash-out of contrast in portovenous phase which is a classical feature of hepatocellular carcinoma in multiphase contrast CT scan

Fig. 46.11: CT scan of abdomen: Contrast-enhanced scan demonstrating a hypodense lesion in right lobe of liver posteriorly due to an abscess

energy to sound energy. These are the active portions of the ultrasonic transducers. **The commonly used substance in the transducer is lead zirconate titanate (PZT).**

Applications

- Ultrasonic beam of **high frequency** gives excellent resolution images of only superficial structures. This is used for study of musculoskeletal system, joints, thyroid, scrotum, etc.
- For imaging **deeper structures of abdomen, a low frequency** probe with greater **penetrancy** is used.

Interpretation of images

- Images are dependent on the **intensity of echoes** received back by the transducer.

Fig. 46.12: Ultrasound of liver and gall bladder demonstrating an isoechoic mass lesion occupying the lumen of gall bladder which was due to malignancy

Fig. 46.13: Ultrasound of liver shows hydatid cyst which is anechoic with multiple daughter cysts

- Structures which reflect all the sound waves back are depicted as **bright echoes** and **termed hyperechoic**.
- Structures which reflect **moderate** level of sound waves appear as uniform grains and are **termed isoechoic**.
- Fluid filled structures which transmit all the sound waves **do not reflect any echoes and are termed hypoechoic**.
- The reflection of sound waves in the form of echoes depends on the density of the organ and the transmission of sound through the same.

Advantages of ultrasonography

1. It is a cost-effective investigation
2. It is widely available
3. Noninvasive
4. Owing to the relatively small size of the apparatus, it is fairly portable, and can thus be brought to the bedside of the moribund patient.
5. It does not involve the use of ionising radiation, and can therefore be safely used in a pregnant patient and can be repeatedly used as a follow-up modality.

Limitations of ultrasonography

1. Its use is limited in thorax
2. Limited use in the abdomen when there is gaseous distension
3. Operator expertise is all important
4. It cannot image bone

MAGNETIC RESONANCE IMAGING (MRI)

Principle

Certain atomic nuclei, which possess unpaired protons or neutrons, have an inherent spin. The nucleus is positively charged and therefore creates a small magnetic field around itself, when it spins. The human body contains in abundance such spinning nuclei in the atoms of hydrogen which is found in water and lipids (Figs 46.14 to 46.18).

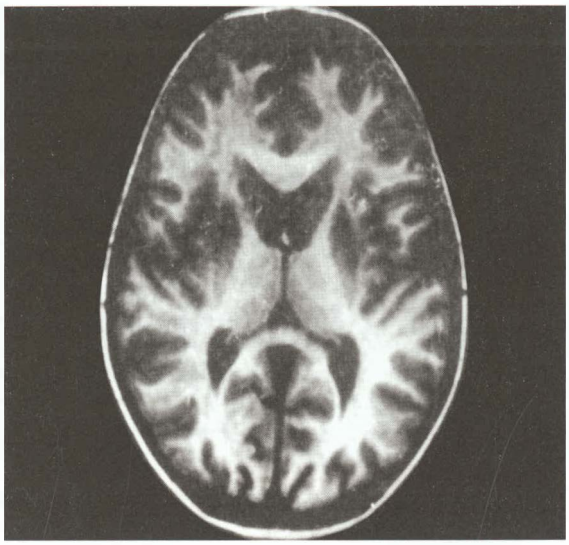


Fig. 46.14: MRI: T1 and T2 weight axial and coronal scans of normal brain

Fig. 46.15: Soft tissue sarcoma involving the muscle of thigh. The lesion is hyperintense in appearance

Fig. 46.16: Carcinoma tongue. MRI image shows hyperintense lesion is involving the posterior and base of the tongue on the left side. The other bright structures seen bilaterally are parotid glands

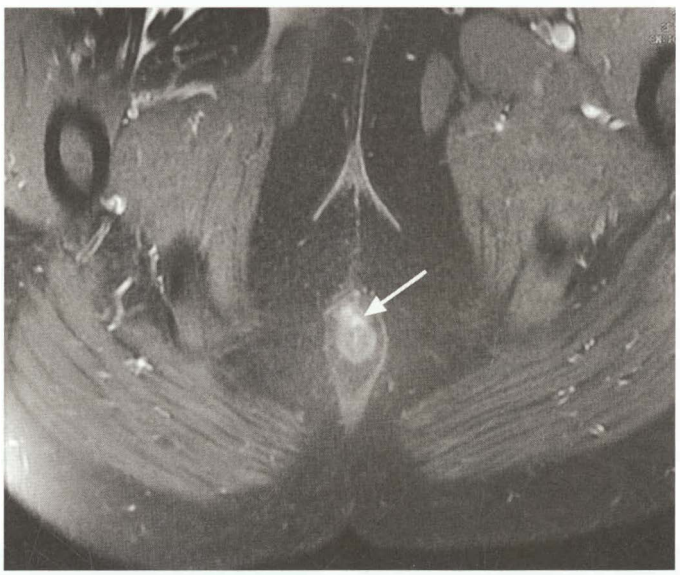


Fig. 46.17: Intersphincteric fistula — axial section of MRI pelvis showing hyperintense fistula (arrow)

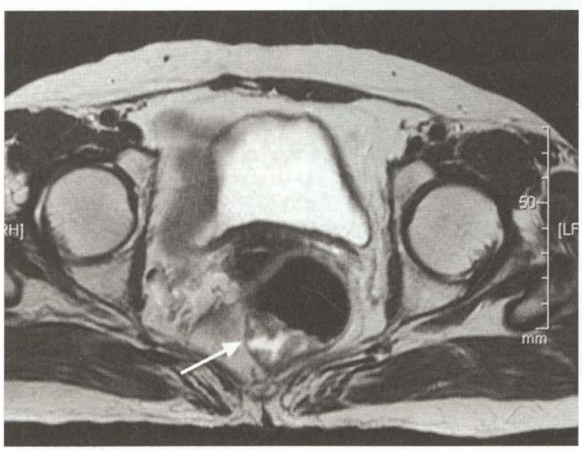


Fig. 46.18: Axial section of pelvis shows wall thickening of the rectal wall (arrow)

When the tissues containing these nuclei are within a strong magnetic field, the nuclei tend to align themselves along the lines of the force. The spinning protons now tend to precess, i.e. wobble about the axis of the main magnetic field. Now a radiofrequency (RF) is applied, being of the same frequency as the processing but at right angles to the main magnetic field. This excites the protons at low energy states into higher energy states. Thus, an absorption of energy takes place, which is used, as the excited protons 'relax' back to their original energy level when the radiofrequency is switched off.

The relaxation of protons back to equilibrium and lower energy state is termed **spin-lattice relaxation** or **longitudinal relaxation**. It is exponential and referred to by the time **constant** T1. When the RF pulse is applied the protons process together in synchronism or in phase with each other. During relaxation, however, they go quickly out of phase due to small variations in local magnetic fields. This loss of phase is termed **spin-spin relaxation** or **transverse relaxation**. It is also an exponential and referred to by the time constant T2. Depending on the type of tissue under study, the T1 and T2 relaxation times will differ, thus giving rise to differences in the image.

The MRI image depends upon 4 main factors:

1. The T1 relaxation time
2. The T2 relaxation time
3. The proton density
4. The blood flow

Depending on the characteristics of the above four parameters, the signal intensity of the image will vary, thus deciding the appearance that any given tissue will finally cast.

Advantages of MRI

1. It is noninvasive
2. It does not involve the use of ionising radiation. Hence, it is safe in that respect.
3. It gives high intrinsic contrast
4. Direct transverse, sagittal and normal imaging possible
5. No bone/air artefact
6. It has no known biological hazard

Disadvantages of MRI

1. The imaging time is long. Hence, movement of the patients may produce artefacts.
2. Due to variety of protocol options during scanning, the final image is highly operator-dependent and this requires expert technical staff.
3. Expensive
4. Poor bone and calcium detail
5. **Patients with pacemakers, metallic implants and critically ill patients cannot be scanned.**

PERIPHERAL VENOGRAPHY

This is the contrast study of the veins of the limbs.

Indications

1. Deep venous thrombosis
2. Demonstration of incompetent perforators
3. In case of suspected venous obstruction by tumour
4. To outline venous malformations.

Procedure

- Tourniquet is applied just above the ankle or elbow t occlude the superficial venous system
- A 19 g butterfly needle is inserted into a distal vein an contrast media (iodine containing) is injected
- Filming is done up to the region of interest

Interpretation of study

- Thrombosis of deep veins are seen as filling defects
- Incompetent perforators are evident as refluxing of contras from deep to superficial system and prominent tortuou collaterals
- Displacement of the opacified veins are seen at sites o tumour. Tumour encasement of veins and thrombosis als may be demonstrable.

Complications

- Complications due to contrast media
- Thrombophlebitis
- Tissue necrosis due to extravasation of contrast
- Pulmonary embolus due to dislodged clot

MYELOGRAPHY (Fig. 46.19)

Definition

This is the study of the spinal canal and spinal cord by injection of contrast medium into the thecal subarachnoid space.

Indications

Suspected intraspinal or nerve root abnormalities

Contraindications

- Papilloedema
- Recent lumbar puncture, since CSF collected outside the thecal space may be tapped.
- Previous adverse reactions to intrathecal injection of contrast medium.

Procedure

- Lumbar puncture is most preferred. The puncture is done at L2–L3 level.

Fig. 46.19: Thoracic myelogram: Contrast in the thecal sac showing a total interruption in the flow of contrast cranially due to an intradural mass lesion

- Once good flow of CSF is obtained, low osmolar nonionic contrast media is injected.
- Filming of lumbar region is done and with adequate tilt towards head end, thoracic and cervical regions are also filmed.
- Cisternal puncture is done when lumbar puncture is a failure.

Interpretation of study

- Disc lesions are seen as impressions on contrast column and cut off the nerve roots at a particular level.
- Intraspinal tumours cause widening of the contrast column with or without total obstruction to flow.
- Vertebral and paravertebral lesions result in displacement of the contrast column and cut off at a particular level.

Complications

- Headache
- Convulsions—rare
- Transient increase in lumbar or sciatic pain
- Hypotension
- Spinal cord damage in cisternal puncture

INTERVENTIONAL RADIOLOGY

The role of radiology was limited as only a diagnostic art until mid 1970s. However, now radiology has taken on an exciting new aspect and has entered the field of interventional radiology. Two main types of interventional procedures: Vascular and nonvascular.

Vascular

1. **Angioplasty:** This is performed by the use of intraluminal balloon catheters and may be performed for almost any diseased vessel in the body. The more commonly treated vessels are the coronaries, renal arteries, peripheral limb vessels, etc.
2. **Embolisation:** This procedure is performed either preoperatively to reduce the vascularity of certain tumours, or as a curative treatment for vascular malformations, aneurysms, GI bleeding, etc. Temporary embolisation may be achieved by using gel foam or autologous clots and permanent embolisation by using balloons, steel coils, ethanol, etc. Inferior vena cava (IVC) umbrella placement, IVC membranotomy are also done.
3. **Intravascular ultrasound:** The use of ultrasound inside a blood vessel to visualise the interior of the vessel in order to detect problems inside the blood vessel.
4. **Stent placement:** A tiny, expandable coil, called a stent, is placed inside a blood vessel at the site of a blockage. The stent is expanded to open up the blockage.
   - **Important types of stents and stent selection:**
     Self-expanding stents are compressed within a catheter device and released by removing a constraining sheath or membrane. The final diameter of the stent is a function of the outward elastic load of the stent and the inward recoil of the elastic wall.
   - **Balloon expandable stents** are mounted on angioplasty balloons in a compressed state and then deployed by balloon inflation. These stents retain the diameter imposed by angioplasty balloon unless externally compressed.
5. **Foreign body extraction:** The use of a catheter inserted into a blood vessel to retrieve a foreign body in the vessel.
6. **Needle biopsy:** A small needle is inserted into the abnormal area in almost any part of the body, guided by imaging techniques, to obtain a tissue biopsy. This type of biopsy can provide a diagnosis without surgical intervention.
7. **Blood clot filters:** A small filter is inserted into a blood clot to catch and break up blood clots.
8. **Injection of clot-lysing agents:** Clot-lysing agents, such as tissue plasminogen activator (tPA) are injected into the body to dissolve blood clots, thereby increasing blood flow to the heart or brain.
9. **Catheter insertions:** A catheter is inserted into large veins for giving chemotherapy drugs, nutritional support, and haemodialysis. A catheter may also be inserted prior to bone marrow transplantation.
10. **Cancer treatment:** Administering cancer medications directly to the tumour site.

Nonvascular

1. **Hepatobiliary:** Percutaneous transhepatic biliary drainage (PTBD) is widely accepted in cases of biliary obstruction, along with percutaneous biliary calculus removal. Biliary stent placement across a malignant lesion is widely being done in inoperable cases as a palliative procedure.
2. **Urinary:** Percutaneous nephrostomy, percutaneous stenting and percutaneous nephrolithotomy are being performed.
3. **Guided biopsy:** Fluoroscopy, ultrasound or CT-guided biopsy of various lesions are now part of routine technique.
4. **Other interventional procedures:** Percutaneous gastrostomy, catheter drainage of abscesses, pseudocysts, ultrasound-guided intrauterine foetal surgeries, etc.

Advantages of interventional procedures

1. Patient compliance is high as surgery is avoided
2. Cost-effectiveness is high

3. Infection rates are low
4. Can be repeated as it is relatively noninvasive
5. Certain untreatable conditions are treated palliatively with interventional procedures.

POSITRON EMISSION TOMOGRAPHY (PET SCAN)

- PET scan is a medical imaging technique that combines computed tomography (CT) and nuclear scanning. It is used to determine the metabolic or biochemical activity in the brain, heart and other organs by tracking the movement and concentration of a radioactive tracer injected into the blood stream.
- A camera records the tracer's signal as it travels through the body and collects information about the organs. A computer then converts the signals into 3D images of the examined organ, which provide a clear view of ar abnormality.
- One of the main differences between PET scan and othe imaging tests like CT or MRI is that the PET scan reveal the cellular level metabolic changes occurring in an orgar and functional changes at cellular level. A PET scan ofter detects these changes very early, whereas CT or MRI detec changes a little later as the disease begins to cause structura changes in organs or tissues.
- Positron emission tomography (PET-CT) constitutes majo progress in management of cancer patients for the initia diagnosis, staging and follow-up of various malignancies PET-CT is also useful in the follow-up of patients following chemotherapy or surgical resection of tumour, since, mos of them have a confusing appearance at CT or MR imaging due to postoperative changes or scar tissue (Figs 46.20 to 46.25).

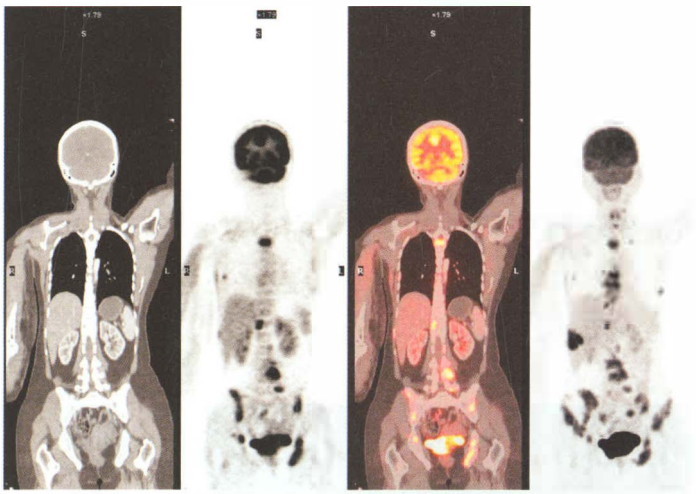


Fig. 46. 20: Carcinoma breast with metastasis

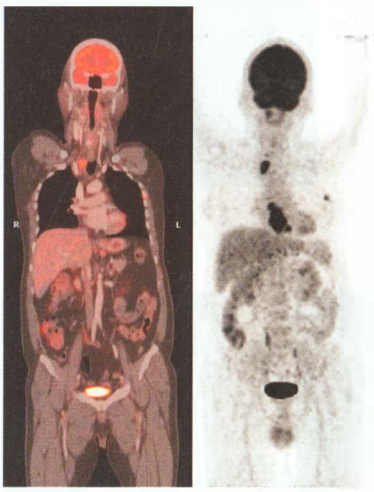


Fig. 46. 21: Carcinoma oesophagus

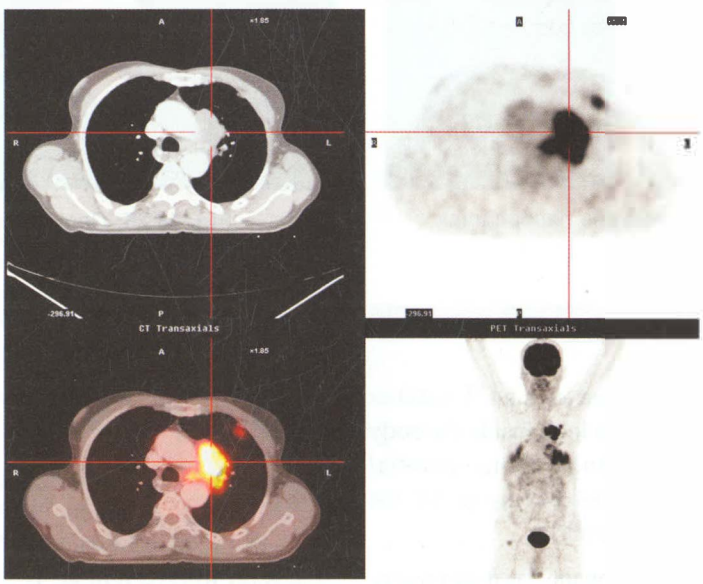


Fig. 46. 22: Carcinoma lung—PET-CT scan

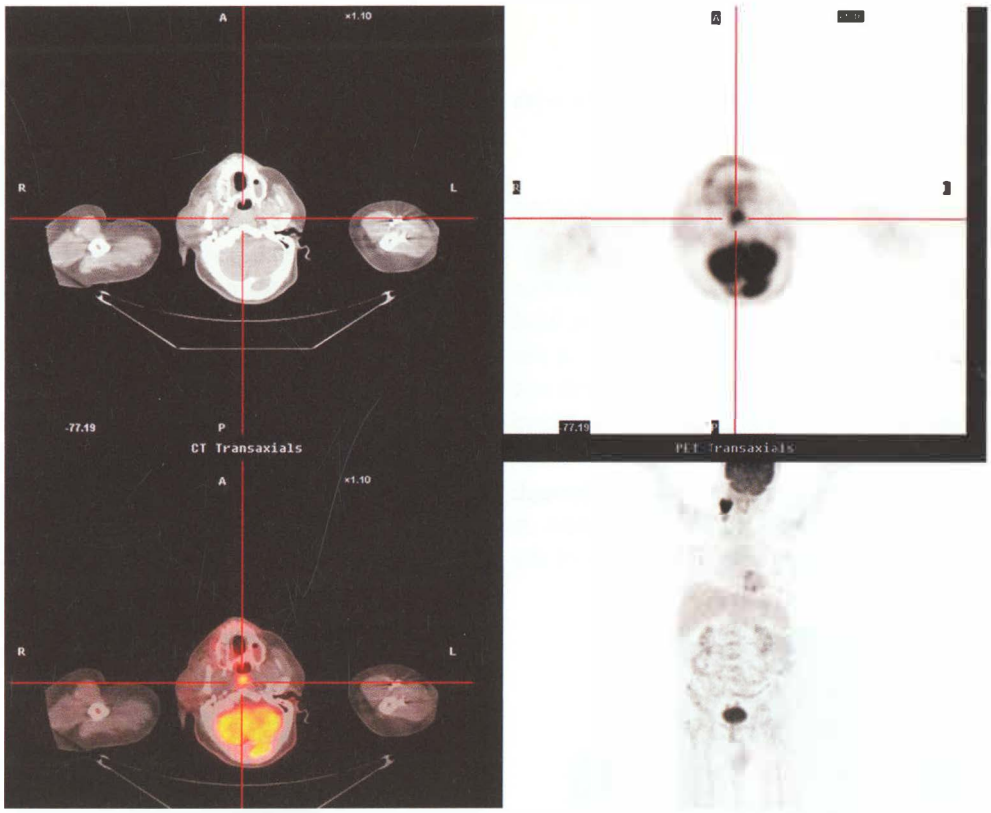


Fig. 46. 23: Carcinoma nasopharynx—PET-CT scan

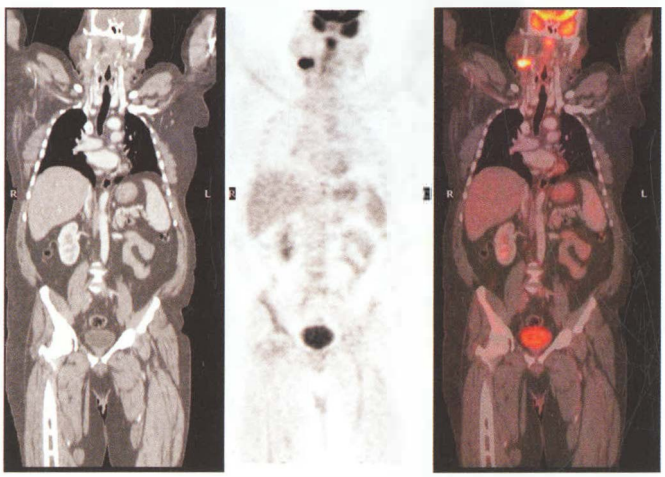


Fig. 46.24: Another case of carcinoma nasopharynx

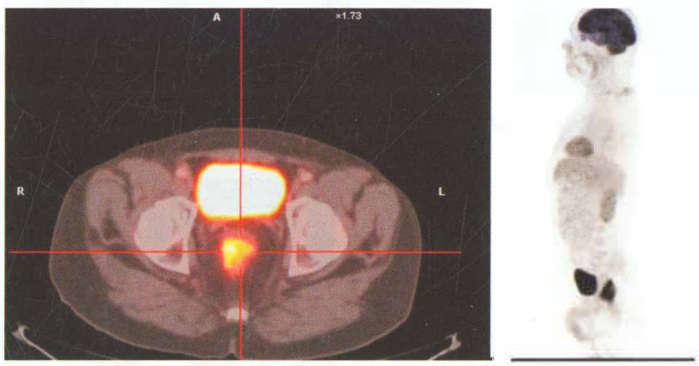


Fig. 46.25: Carcinoma rectum—PET-CT scan

VIRTUAL COLONOSCOPY

- It is a recently developed technique that uses a CT scanner and computer virtual reality software to look inside the body without having to insert a long tube (conventional colonoscopy) into the colon or without having to fill the colon with liquid barium (barium enema).
- More formally known as three-dimensional CT colonography, the virtual procedure allows radiologists to obtain 3D images from different angles, providing a sort of movie of the colon's interior without having to insert an endoscope into the bowel.

Advantages

- Noninvasive procedure, well tolerated by patient
- Requires no sedation, less time-consuming
- Useful in elderly who are frail and infirm
- Useful when a tumour is large enough to block passage of scope (Figs 46.26 and 46.27).

Figs 46.26 and 46.27: Virtual colonoscopy shows opened up view to look for small mass or polyp

Disadvantages

- Exposure to radiation, less detail of inner lining of colon
- Small polyps are located more reliably by colonoscopy
- Strictly a diagnostic procedure (unlike colonoscopy).

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated.
- PET scan, virtual colonoscopy have been upgraded
- A few photographs have been added

MULTIPLE CHOICE QUESTIONS

1. Niche and a notch is diagnostic of:
A. Chronic duodenal ulcer
B. Chronic gastric ulcer
C. Carcinoma stomach
D. Stromal tumour

2. Trifoliate/Clover deformity is diagnostic of:
A. Chronic duodenal ulcer
B. Chronic gastric ulcer
C. Carcinoma stomach
D. Stromal tumour

3. The substance used in barium studies is:
A. Barium chloride B. Barium sulphate
C. Barium carbonate D. Barium sulphide

4. Barium follow through extends up to:
A. Proximal duodenum
B. Fundus of the stomach
C. Ileocaecal junction
D. Proximal jejunum

5. Which of the following is a contraindication for barium study?
A. Dysphagia and odynophagia
B. Motility disorders of the GIT
C. Perforation of gastric mucosa
D. Assessing mediastinal masses

6. Achalasia cardia shows the following findings on barium swallow *except*:
A. Shouldering effect cranial and caudal to the lesion
B. Rat tail appearance of lower end of the oesophagus
C. Gross dilatation of the proximal oesophagus
D. Thin streaks of contrast entering the stomach

7. 'Moulage' sign (barium in a featureless tube) is a feature of:
A. Crohn's disease B. Intestinal TB
C. Ulcerative colitis D. Malabsorption

8. Which of the following statements is false?
A. Angiographic studies use ^{123}I as contrast substance
B. Enteroclysis is done for mechanical obstruction of the intestine
C. Peripheral venography done for deep vein thrombosis uses ^{125}I
D. Papilloedema is an absolute contraindication for myelography

9. Which of the following statements is false?
A. Ultrasound is based on the principle of piezoelectric effect, the most common substance used in ultrasonic transducers being lead zirconate titanate (PZT)
B. MRI uses ionising radiation, hence it is unsafe
C. Patients with pacemakers and critically ill patients cannot be scanned using MRI
D. CT scan gives good bone and calcium detail

10. The most common side effect of peripheral venography is:

A. Complications due to contrast
B. Tissue necrosis
C. Thrombophlebitis
D. Pulmonary embolism due to dislodged clot

11. Which of the following statements is false about PET scan?

A. PET scan uses protons for radiological examination
B. It combines CT and nuclear scanning
C. It detects changes at the cellular level
D. It helps in early detection of changes in various pathologies.

12. BI-RADS score of 5 indicates which of the following?

A. Normal mammogram, no evidence of cancer
B. Mammogram normal, some evidence of cancer present
C. Suspicious findings on mammogram, 20–35% chance of cancer
D. Mammogram findings highly suspicious, 95% chance of cancer

13. The following statements about virtual colonoscopy ar true *except*:

A. It is also called CT pneumocolon, purely diagnostic
B. It is invasive, requires sedation and contraindicated i elderly
C. It involves exposure to radiation
D. It cannot identify polyps measuring betwee 2 mm and 10 mm

14. Which of the following in future may be a gold standard test for screening of colorectal cancer:

A. PET scan
B. Sigmoidoscopy and colonoscopy
C. MRI
D. Virtual colonoscopy

15. The following is true about MRI *except*:

A. It is noninvasive
B. Gives high intrinsic contrast
C. Imaging possible in transverse, sagittal and normal views
D. Bone/air artefact can be a problem

ANSWERS

1 B	2 A	3 B	4 C	5 C	6 A	7 D	8 A	9 B	10 A
11 A	12 D	13 B	14 D	15 D					

47

Principles of Clinical Radiation Oncology

- Radiation
- Dose fractionation
- Sources and methods
- Measurement
- Clinical uses
- Curative treatment
- Palliative treatment
- Radiotherapy reactions
- Advances in radiation therapy
- What is new?/Recent advances

Introduction

Radiation oncology is that discipline of human medicine concerned with the generation, conservation, and dissemination of knowledge concerning the causes, prevention, and treatment of cancer and other diseases involving special expertise in the therapeutic applications of ionising radiation.

Leopold Freund was the first to report in 1897, the use of ionising radiation to "cure" a large nevus pigmentosus on the back of a young girl. With time, ionising radiation became more precise; high-energy photons, electrons, protons, neutrons, and carbon ions became available; and treatment planning and delivery became more accurate and reproducible. Advances in computer and electronic technology fostered the development of more sophisticated treatment-planning and delivery techniques, leading to the development and eventually broad implementation of three-dimensional conformal radiation therapy (3DCRT) and intensity-modulated radiation therapy (IMRT), stereotactic body radiation therapy (SBRT), etc.

RADIATION

The term radiation applies to the emission and propagation of **energy** through space and material medium. Radiation travels with the speed of light in a vacuum, and interacts with living or nonliving matter resulting in varying degrees of energy transfer to the biological medium. This process of deposition of energy within the cells is brought about by **ionisation** (removal of an orbital electron) of atoms and molecules. Ionising radiations are off very high frequency (3×10^{21} hertz) and short wavelength (10^{13} m) electromagnetic waves. Ionisation can occur within the nuclear DNA molecule of a cell (**directly acting**) or interaction with other molecules, mainly water (H_2O) to produce **free radicals (indirectly)**, which in turn can damage DNA and result in cell death or mutagenesis.

By damaging DNA, radiation interferes with cell division and can result in **reproductive** death of a cell. This process in a malignant tumour could mean the loss of its ability for uncontrolled cell division or proliferation. This process is unselective; it occurs both in cells of normal tissues and in those of tumours. Therapeutic usefulness of radiotherapy, therefore, depends on the differential sensitivity of tissues (normal *vs* tumour cell), on careful treatment planning and dose prescription to minimise normal tissue damage and the patient's tolerance to radiation.

DOSE FRACTIONATION

The five **Rs** of radiobiology provide the basis for fractional radiotherapy (Key Box 47.1). In clinical practice dividing a dose into a number of fractions has the following advantages:

1. The **acute effects** of single doses of radiation can be decreased with fractionation. The patient's symptomatic tolerance improves with **fractional radiation.**
2. Fractionation exploits the difference in **recovery rate** between normal tissues and tumours. Effects on normal tissues are less because of repair of sublethal damage between dose fractions and normal cellular **repopulation.**

3. Radiation-induced **redistribution** of cells within the cell cycle tends to sensitise the rapidly proliferating cells, which is seen more in tumours.
4. **Radiosensitivity** of cells depend markedly on the phase of the cell cycle at which they receive the radiation. Cells in **mitosis and G2 phase are the most sensitive** and cells in early Gl, and late S phases are the most resistant.
5. Reduction in the number of hypoxic cells is brought about through cell kill and **reoxygenation**. Also blood vessels compressed by a growing cancer are decompressed as the cancer shrinks, permitting better oxygenation.

KEY BOX 47.1

BASIS OF FRACTIONAL RADIOTHERAPY

- Repair
- Redistribution
- Reoxygenation
- Repopulation/recovery
- Radiosensitivity

Oxygen effect

The biologic effects of ionising radiations are greatly influenced by the presence of normal oxygen concentration within the cells. The absence or low oxygen content conveys a resistance to radiation requiring about three times the dose to produce the same biologic effects. Certain solid tumours and large tumours are likely to contain 10–15% of hypoxic cells.

For routine practice the "Conventional or Standard" dose fractionation schedule is followed. This consists of 180–200 cGy fraction per day, and 5 fractions per week over 4–6 weeks (depending on the total dose). The choice of optimal dose/time/fractionation schedules for various tumours should be individualised according to the cell kinetic characteristics and clinical observations.

Radiocurability refers to the eradication of tumour at the primary or regional site and reflects the direct effect of irradiation, which may or may not parallel the patient's ultimate outcome.

PROBABILITY OF TUMOUR CONTROL (Fig. 47.1)

It is axiomatic in radiation therapy that higher doses of radiation produce better tumour control, and numerous dose-response curves (sigmoid in shape) in a variety of tumours have been published.

For every increment of dose a certain fraction of cells will be killed. Therefore the total number of surviving cells will be proportional to the initial number present and the fraction killed with each dose. Thus, it is apparent that **various levels of irradiation yield a different probability of tumour control**, depending on the extent of lesion or number of clonogenic cells present.

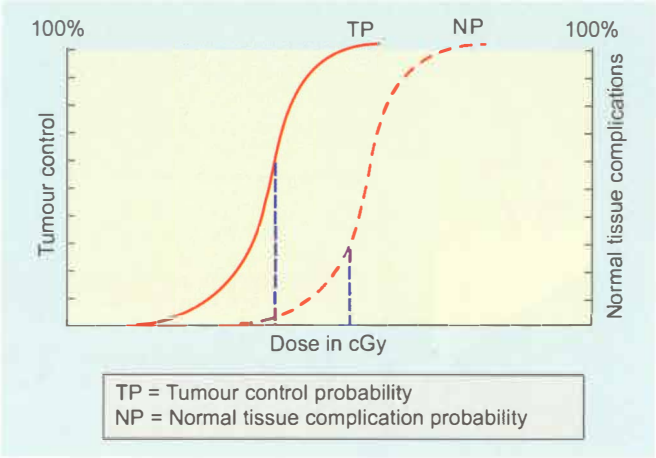


Fig. 47.1: The graph showing tumour probability control

For **subclinical disease** (10^{3-4} cells) in squamous cell carcinoma of the upper respiratory tract or for adenocarcinoma of the breast, doses of 4500–5000 cGy result in control of disease in over 90% of patients.

For **microscopic residual disease** or cell aggregates greater than $10^6/10^9$ are required for the pathologist to detect them. Therefore, these volumes must receive higher doses of radiation in the range of 6000–6500 cGy in 6–7 weeks for epithelial tumours.

For **clinically palpable tumours (Gross disease)**, doses of 6000 cGy (for Tl) to 7500 cGy to 8000 cGy for T4 tumours) are required (200 cGy/day/5 fractions weekly). This dose range and probability of tumour control have been documented for various tumours.

RADIOTHERAPY—SOURCES AND METHODS OF DELIVERY

Radiotherapy is the therapeutic use of high-energy ionising radiation in the treatment/management of malignant disease.

These are either electromagnetic waves, X-rays, gamma rays or corpuscular (subatomic particles) electrons, protons, neutrons, alpha particles, or heavy ion nuclei. Ionising radiation penetrates tissues to different depths according to its type of energy and physical nature. Radiotherapy treatment planning is an important part of the radiation oncologist's work.

SOURCE OF RADIATION (Fig. 47.2)

Gamma and beta rays from radioactive isotopes (cobalt 60, caesium 137, indium 197) and X-rays and electrons from a high energy X-ray machine (linear accelerator). Protons, neutrons and heavy ion nuclei from cyclotrons.

X-rays and gamma rays are identical in properties but are produced by different sources. Ionising radiations can be classified according to their density of ionisations per unit length of the distance in the absorbing media as low and high LET (linear energy transfer) radiation.

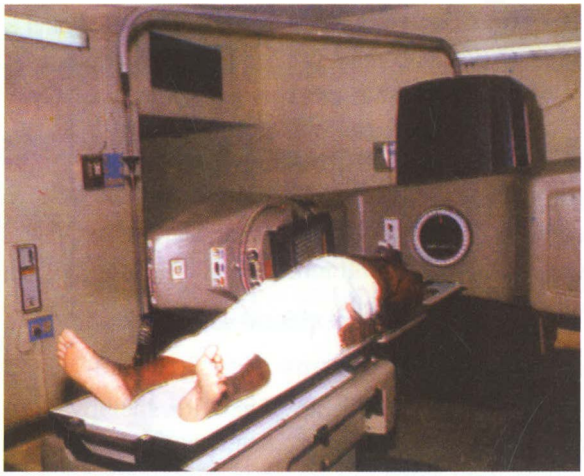


Fig. 47.2: Cobalt teletherapy unit

IONISING RADIATION

- Low LET—X-rays, gamma rays and electron.
- High LET—neutrons, protons, α-particles and negative ions.
- High LET radiation has a mass heavier than electrons. Hence, they cause dense ionisation and are biologically more effective (damaging) than low LET radiation. They are also less dependent on repair of sublethal damage, cell cycle phases and oxygen content of the cells for radio-sensitivity.
- High LET radiation is available in limited cancer centres around the world, require expensive equipment to produce and are undergoing clinical trials.

METHODS OF DELIVERY

Ionising radiations may be delivered clinically in three ways:

I. External beam irradiation

From sources at a distance (usually 80–100 cm) from the body surface. This includes ^{60}Co Teletherapy Units and X-ray sources, such as linear accelerators (Fig. 47.3).

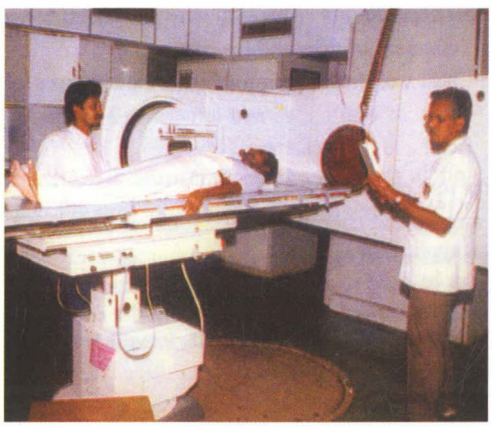


Fig. 47.3: Linear accelerator

Advantages of megavoltage beam RT

1. Deeper penetration
2. Sharp beam edges
3. Skin sparing
4. Equal absorption in bone and soft tissues
5. Improved dose distribution within tissue.

II. Brachytherapy (Key Box 47.2)

Brachytherapy refers to use of radiation sources in or close to the tumour.

Use of sealed (closed containers) radioactive sources for radiation treatment from a short distance.

Types of brachytherapy

1. **Intracavitary** (within a cavity), e.g. uterine cavity, vagina cavity, oesophageal and bronchial lumen.
2. **Interstitial** when radioactive needles and wires are inserted into and around a tumour.
3. **Surface moulds or plaques** as radioactive surface applicators, e.g. skin cancer, eye cancers.

With this mode of therapy a high dose can be delivered locally to the tumour with rapid dose fall off in the surrounding normal tissue. In the past, brachytherapy was carried out mostly with radium or radon sources. Currently use of artificially produced radionuclides such as ^{137}Cs (caesium 137) , ^{192}Ir (iridium 192), ^{198}Au (gold 198), and ^{125}I (iodine 125) is rapidly increasing.

New technical developments have stimulated increased interest in brachytherapy

1. Introduction of improved artificial isotopes
2. Manual afterloading devices to reduce personnel exposure
3. Remote afterloading, high dose rate (HDR) machines, have increased accuracy, improved dosimetry, reduced (short) treatment time, treatment on outpatient basis and have improved patient compliance.

Brachytherapy is used very often. **Combined** with **external beam treatment** and **rarely alone** (early stage). The rationale behind combining the two is to treat the primary site and regional spread (very often subclinical disease) with external RT and to deliver a higher dose (boost) to the primary (gross disease) with brachytherapy. Aim is not to exceed the normal tissue tolerance

KEY BOX 47.2

ADVANTAGES OF BRACHYTHERAPY

1. High localised dose to limited volume
2. Minimal dose to adjacent tissues
3. Spares deeper normal tissues
4. Continuous RT in a single course
5. Short overall time (high dose rate)
6. Alone: Rarely (early stage)
   Combined: Often (as boost)

and at the same time the tumour should receive adequate curative dose. Brachytherapy can also be used as palliative therapy, e.g. bronchial and oesophageal obstruction.

III. Internal or systemic irradiation

From unsealed radioactive sources (i.e. ^{131}I, ^{32}P, ^{89}Sr) administered enterally, intracavitarily or intravenously, for diagnostic (nuclear medicine) and therapeutic purposes, e.g. carcinoma thyroid, bone tumours and thyrotoxicosis.

MEASUREMENT OF IONISING RADIATION

1. The **Roentgen** is a unit of exposure. It is a measure of ionisations produced per unit volume of air by X-rays and gamma rays and cannot be used for photon energies above 3 Mev.
   - The SI unit for exposure is Coulomb per kilogram (C/kg)
   - 1 R = 2.58×10^4 C/kg air
2. **Radiation absorbed dose (RAD)**
   - Absorbed dose is a measure of the biologically significant effects produced by ionising radiation. Absorbed dose = De/dm, De is the mean energy imparted by the ionising radiation to material of mass dm. The old unit of dose is rad and represents the absorption of 100 ergs of energy per gram of absorbing material.
   - 1 rad == 100 ergs/g = 10^2 J/kg
   - The SI unit of absorbed dose is gray (Gy) and is defined as
   - 1 Gy = 1 J/kg

Thus the relationship between rad and gray is 1 Gy = 100 rad or 1 cGy = 1 rad.

ELECTRON BEAM THERAPY

Source

Mainly linear accelerators

Energy

Most useful range for clinical use is 6 to 20 Mev.

Use

1. Superficial tumours up to a depth of 5 cm
2. Local boost

Advantages

1. Characteristic sharp dose fall off beyond the tumour
2. Dose uniformity within the target volume

Principal applications

1. Treatment of skin and lip cancers
2. Chest wall irradiation for breast cancer
3. Boost dose to nodes and tumour bed
4. Head and neck cancers.

Clinical use of radiotherapy (Key Box 47.3)

Like surgery and chemotherapy, radiation therapy (RT) has definite indications and contraindications for its application.

It can be used alone to cure or, in combination with other methods, as an adjuvant. Currently 50–60% of all patients with cancer receive RT during the course of their illness. If properly used, 50% of these patients could get cured. For the other half, incurable by any current method, palliation of specific symptoms and signs can improve quality of life (Table 47.1).

Before treating a patient with radiotherapy, the radiotherapist must be satisfied that the working diagnosis is correct, pretreatment investigations and staging have to be complete. Then the radiation oncologist must address two questions:

1. Is the treatment intent ***curative*** or ***palliative***?
2. What is the best approach to achieve this goal?

The first question is vital, for there are important differences between radical and palliative radiotherapy.

The second question recognises that cancer can be treated by surgery, radiotherapy and drugs. In many instances, radiotherapy is the best approach. In view of the increasing complexity of curative cancer management for many tumours, with different combinations of surgery, radiotherapy and chemotherapy for different stages of the disease, require a co-ordinated **Multidisciplinary** approach. The correct initial choice gives the best prospect for cure or good palliation.

AS A PRIMARY CURATIVE MODALITY (Key Box 47.4)

A. RT frequently may be the sole agent used with curative intent for anatomically limited tumours of the retina, optic nerve, brain (craniopharyngioma, medulloblastoma,

Table 47.1 Differences between radical and palliative radiotherapy

Radical radiotherapy	Palliative radiotherapy
• Treatment is intended to eradicate all clonogenic malignant cells.	• Treatment is intended to control symptoms to improve quality of life.
• High dose curative courses of RT needed and considerable normal tissue morbidity (acute) may be associated.	• Minimum doses of RT to achieve maximum control with minimal side-effects.
• Late side-effects of adjacent normal tissues may be dose limiting.	• Short patient survival times, less concern with long-term limiting, morbidity.
• Patient often needs high doses and long courses of treatment.	• Patient prefers few hospital visits, so short courses of treatment used.

KEY BOX 47.3

RADIOTHERAPY USE IN FOUR SETTINGS

1. As a primary **curative** modality
2. As an **adjuvant** for curative therapy (combined modality)
3. As **prophylactic radiation**
4. As **palliative** treatment

KEY BOX 47.4

INDICATIONS FOR CURATIVE RT

Stages I and II

Testis	Seminoma
Ovary	Dysgerminoma
Skin	Basal cell and squamous cell carcinoma
Lymphatic	Hodgkin's lymphoma
Cervix	Cervix, uterus, vagina
Bladder	Transitional cell Ca
Prostate	Adenocarcinoma
Anal canal	Carcinoma
Head and neck	Cancers
Oesophagus	Cancer
Lung	Non-small cell cancer
Brain	Medulloblastoma

ependymoma), spinal cord (low-grade glioma), skin, oral cavity, pharynx, larynx, oesophagus, uterine cervix, vagina, prostate and reticuloendothelial system (Hodgkin's disease, stages I, II and III A).

B. When no other potentially curative treatment exists. Some cancers remain localised for all or much of their natural history. These cancers might also be unresectable by virtue of their anatomical location or because of local infiltration into surrounding normal/vital structures, which would mean that surgery will severely affect physiological function, e.g. locally advanced head and neck cancer, cervical cancers stage IIb-III, medulloblastoma (alternative to surgery for inaccessible and inoperable malignancies).

C. Where alternative therapy is considered more "toxic". Carcinoma of the larynx, anal canal, breast can all be managed by ablative surgery and in each case the anatomy and physiology of the respective organ is lost. Each of these cancers can be managed by irradiation with preservation of anatomy and function.

Preservation of organ and its function (larynx, breast, anal canal, limbs, cervix, tongue, bladder)

ADJUVANT FOR CURATIVE THERAPY (COMBINED MODALITY) (Key Box 47.5)

RT is combined with surgery for advanced cancers of the head and neck, cancers of the lung, uterus, breast, urinary bladder, testis (seminoma), rectum, soft tissue sarcomas and primary bone tumours.

KEY BOX 47.5

ADJUVANT RT (SURG+RT+/–CT)

- Head and neck cancer locally advanced
- Brain tumours
- Breast cancer
- Rectal cancer
- Soft tissue sarcoma
- Bone sarcoma
- Endometrial Ca
- Paediatric solid tumours (Wilms', rhabdomyosarcoma, neuroblastoma)

RT is an adjuvant to chemotherapy for some patients with lymphomas, lung cancers and cancer in children (rhabdomyosarcoma, Wilms' tumour, neuroblastoma).

In some clinical situations the combined benefits of surgery RT and chemotherapy might be exploited. It has been most useful in the management of breast cancer, bone sarcoma and Wilms' tumour.

COMBINED TREATMENT (SURGERY AND RADIATION THERAPY)

In many situations radiation therapy alone is inadequate for achieving maximum cure levels. This can be because the number of tumour stem cells is too large, some or all of the cancer cells are radioresistant, or tolerance of the contiguous normal tissues is too low. The rationale for combining surgery and radiation therapy is the differing mechanisms of the two disciplines. Radiation therapy fails at the centre of the tumour where the concentrations of the tumour cells is the largest and the conditions may be hypoxic (less sensitive to RT).

Surgical resection fails because the tumour extends further than the margins of excision, infesting contiguous tissues with undetectable microscopic foci. Radiation therapy is efficient in the sterilisation of these tumour cell numbers that are well vascularised, and the surgical resection is efficient in removing the gross necrotic tumour masses.

Radiation can be combined with surgery either preoperatively or postoperatively.

Aims and advantages of preoperative RT

1. Unresectable cancer to resectable cancer
2. Prevent iatrogenic metastases
3. Reduction of size and vascularity
4. Destroys microscopic foci beyond surgical margin

Disadvantages

1. Delay in surgical (primary) treatment
2. Delay in wound healing
3. Pathologic downstaging to influence other adjuvant treatment.

4. Alters anatomical staging (precise pathological extent)
5. Inability to tailor RT to high risk areas.

Postoperative RT (Key Box 47.6)

KEY BOX 47.6

AIM AND ADVANTAGES

1. Exact disease extent known so as to tailor the treatment individually.
2. Operative margins are well-defined—gross or microscopic.
3. Less postoperative complications—wound healing intact.
4. GI anastomoses and ileal conduits can be done in a non-irradiated field.
5. Potential for unnecessary irradiation in some patients is reduced

DISADVANTAGES

1. Delay in RT due to postoperative complications—delay wound healing.
2. Decreased radiosensitivity of the tumour due to rich oxygen in vascular tumour.
3. Postoperative adhesions of organs/structures increase RT complication rate.
4. No effect on dissemination during surgery.
5. Volume of normal tissue to be irradiated is more. Usually all tissue planes are potentially contaminated by surgery.

Clinical situations may indicate different sequences but such combinations of surgery and radiation therapy improve the local tumour control rate for many advanced cancers. Combined therapy may also improve the cure rate at the same time reducing the morbidity associated with more aggressive single modality treatment.

COMBINATION OF RADIOTHERAPY WITH CHEMOTHERAPY

In general, chemotherapy is used in an adjuvant way to control subclinical disease elsewhere in the body or in an additive way to enhance the local effects of the radiation to achieve higher rate of local control. Many other agents will act in both ways.

Agents of choice are those whose toxic effects are in organs not included in the radiation target volume. An example is the combination of the cisplatin compounds with radiation therapy for head and neck cancers. Here, the toxicity of chemotherapy is primarily haematogenous and renal. The toxicity of RT is on the oral mucosa.

Chemotherapy could be combined with RT in three main ways

1. Neoadjuvant: 1–3 courses before definitive RT.
2. Adjuvant: After completion of definitive RT.
3. Concurrent: During a course of radiotherapy.
4. Combinations of the above.

PROPHYLACTIC CRANIAL RADIATION

Certain cancers have a high incidence of developing brain (CNS) metastases even after their primary disease is controlled since the CNS because of the blood-brain barrier can act as a sanctuary site for relapse. Among such patients, it is possible to reduce their local CNS relapse rate and improve survival by treating the CNS by prophylactic cranial RT ± intrathecal chemotherapy. The total dose needed is low (18–24 Gy) and has minimal side effects, e.g. acute lymphoblastic leukaemia/high grade lymphomas.

KEY BOX 47.7

PALLIATIVE TREATMENT

Objectives of palliative irradiation include:

- Relief of pain, usually from metastases to bone
- Relief of headache and neurological dysfunction from intracranial metastases
- Relief of obstruction, such as tumours involving ureter, oesophagus, bronchus, lymphatic and blood vessels.
- Promotion of healing of surface wounds by local tumour control.
- Haemostatic for cervical/bladder cancers.

MANAGEMENT: RADIOTHERAPY REACTIONS (Table 47.2)

The incidence of systemic symptoms from radiotherapy is variable. In broad terms, the larger the treatment yield, the fraction size and the total given dose, the greater will be the chance of the patient developing problems. The dose of radiation that can be delivered is limited by acute reactions and by late irreversible organ/tissue damage. Each organ has a known tolerance which should not be exceeded.

However, in order to achieve a given level of tumour control probability certain amount of normal tissue sequelae are unavoidable.

During a course of radiotherapy mild to moderate grade acute reactions occur frequently and can be usually conservatively managed and might require a short break in the treatment whereas chronic reactions are usually the dose limiting complications. Severe grade reactions should be avoided using proper time dose fractionation regimens, accurate treatment planning and execution.

Some of the important acute reactions following RT, their threshold doses and management are shown in Table 47.2.

Table 47.2 Complications and management of RT

Organ toxicity		Approx. dose threshold Gy	Specific management points
Skin	Erythema	10–12	No specific treatment required
Skin	Dry desquamation	40–50	Proflavine and emollients may produce symptomatic relief
Shin	Moist desquamation	45–55	Keep the affected area dry. Gentian violet may be helpful in drying the affected area
Mucous	Mucositis	30–40	Topical benzydamine hydrochloride mouth rinse or spray. Stop smoking. Mucaine for oesophageal mucositis. Always exclude candidiasis
Hair	Alopecia	30–40	Warm patient prior to starting treatment. Advise a wig fitting
Lung	Pneumonitis with cough, dyspnoea	20	Consider systemic corticosteroids
GI tract	Nausea, vomiting	Any dose	Regular antiemetics may be required
Gl tract	Diarrhoea	30–40	Advise low fibre diet when starting treatment. Antidiarrhoeal preparations may be required for symptomatic relief
Bladder	Urinary frequency and dysuria	40–50	Exclude urinary tract infection and consider antimuscarinic drugs
Bone marrow	Suppression especially of white blood cells and platelets during wide-field radiotherapy	10–20	Check full blood count regularly

ADVANCES IN RADIATION THERAPY

Current research in radiation oncology is of such significance that it promises a new standard of care for patients with cancer. Recent advances in radiation therapy include efforts to improve the effectiveness of radiation and to improve the quality of life of treated patients. Innovations in radiobiology, imaging technology, computer technology and treatment machine technology has resulted in marked changes in the way radiotherapy is practised at present. The newer methods aim at increasing the accuracy of treatment, planning and dose delivery using the highly sophisticated features offered by the modern day equipment (Key Box 47.8).

KEY BOX 47.8

ADVANCES IN RADIATION THERAPY

1. 3D conformal radiotherapy (3-dimensional)
2. IMRT (intensity modulated radiation therapy)
3. SRS/SRT (stereotactic radiosurgery and radiotherapy)
4. IGRT (image guided radiation therapy)
5. SBRT (stereotactic body radiation therapy)
6. Proton therapy
7. Intravascular brachytherapy
8. CyberKnife

1. **3-D conformal radiotherapy** (three-dimensional treatment planning and conformal dose delivery). In 3-D CRT, patient immobilisation, image-guided treatment planning and computer-controlled treatment delivery can create a radiation dose distribution that conforms to the shape of the tumour volume (Key Box 47.9). The tumour volume containing the cancer and areas of potential cancer is much more accurately outlined as are normal tissues to be avoided. The target radiation dose can be increased when necessary without increased toxicity to normal tissue. This is accomplished using volumetric CT data in a 3-D treatment planning computer.
2. **IMRT:** IMRT is an advanced form of 3-DCRT. It is one of the technologically most advanced treatment methods available in external beam radiation therapy. IMRT allows very precise external beam radiotherapy treatments. Rather than having a single large radiation beam pass through the body, with IMRT the radiation is effectively broken up into thousands of tiny pencil-thin radiation beams of varying

KEY BOX 47.9

CANCERS BEING TREATED WITH IMRT

- Prostate cancer, pancreatic tumours, lung cancer
- Metastatic brain tumours, primary brain tumours (glioblastomas, gliomas, etc)
- Liver tumours (metastases, hepatocellular carcinoma)
- Head and neck cancer (larynx, tongue, sinus, base of skull, mouth, etc)
- Radiosurgery (single fraction) and stereotactic radiation therapy (fractionated)

intensity with millimetre accuracy. These beams enter the body from many angles and intersect on the cancer. This results in a high dosage to the tumour and a lower dose to the surrounding healthy tissues.

- **Intensity modulation radiotherapy** can allow us to treat tumours to a higher dose, retreat cancers that have previously been irradiated, and safely treat tumours that are located very close to delicate organs like the eye, spinal cord, or rectum. Simply put, this can translate into a higher cancer control rate and a lower rate of side effects.

3. SRS/SRT (stereotactic radiosurgery and radiotherapy)

High-dose highly focused radiation therapy for small (SRS = Single fraction, SRT = Multiple fractions) target lesions (< 2-4 cm) can be accomplished by either gamma knife (multiple, fixed, precisely aimed cobalt teletherapy beams) or stereotactic radiation therapy (multiple rotational arcs of photon beams from a linear accelerator). Both techniques are similar in their use of standard energy photon beams for treatment and rely on meticulous patient immobilisation to deliver treatment to a precisely localised target within a co-ordinate mapping system. These techniques have been widely used and well described for the treatment of intracranial neoplasms (meningiomas, acoustic neuromas and metastatic tumours) and for the ablation of arteriovenous malformations and ocular melanomas. [Equipment used: 1. Gamma Knife (Multiple ^{60}Co sources) or 2. X- knife (modified linear accelerator)]

4. IGRT

- Image guided radiation therapy (IGRT), is one of the most cutting-edge innovations in cancer technology available. Tumors can move, because of breathing and other movement in the body. Real-time imaging of the treatment target and normal organs during each treatment allows for minimisation of reduction of irradiated volumes, as well decreases the chance of missing a target, helping to limit radiation exposure to healthy tissue and reduce common radiation side effects.
- In IGRT, the linear accelerators are equipped with imaging technology that take pictures of the tumour immediately before or even during the time radiation is delivered.

  There are numerous types of imaging modalities that can be incorporated into an IGRT system. Examples include: ultrasound images, low-energy (kV) CT scan images, high-energy (MV) CT scan images, etc.

5. SBRT

Stereotactic body radiation therapy (SBRT) is a technique that utilises precisely targeted radiation to a tumour while minimising radiation to adjacent normal tissue (Fig. 47.4). This targeting allows treatment of small or moderate sized

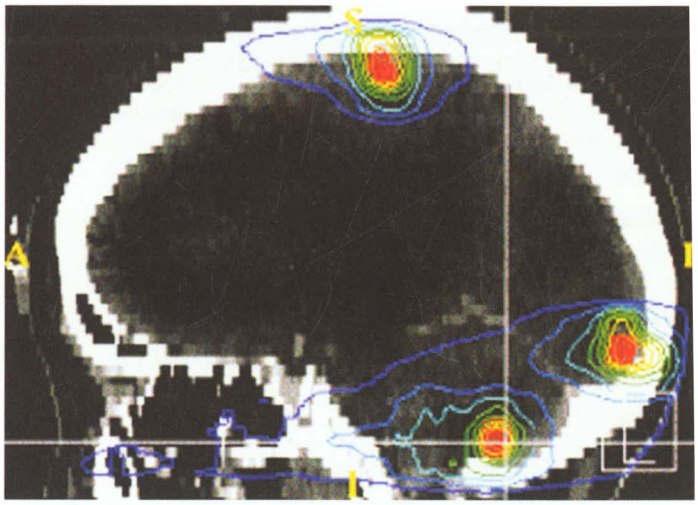


Fig. 47.4: Stereotactic radiosurgery brain (metastasis)

tumours in either a single or limited number of dose fractions. SBRT has been used in hepatocellular carcinoma, lung cancer, prostate cancer, pancreatic cancer. As with any form of radiation therapy, careful attention to matters of patient selection and technical quality assurance is essential for the effective and safe implementation of SBRT.

6. Proton therapy

- **Proton beam:** Proton radiation reduces the dose to normal tissues by allowing for more precise dose delivery because of the unique physical properties of heavy particles. Protons penetrate tissue to a variable depth, depending upon their energy, and then deposit that energy in the tissue in a sharp peak, known as a Bragg peak. This rapid dose fall off at a depth that can be controlled by the initial energy of the protons allows for decreased radiation to adjoining normal tissue by a factor of 2 to 3.
- Protons are used for uveal melanoma (ocular tumours), skull base and paraspinal tumors (chondrosarcoma and chordoma), and unresectable sarcomas, pediatric neoplasms (such as medulloblastoma) and prostate cancer.

7. Intravascular brachytherapy: **Arterial** renarrowing after angioplasty or restenosis occurs in 30 to 40 percent of patients and results from neointimal proliferation and constrictive remodelling of the angio-injured artery. Coronary stenting has led to a 30 to 50 percent decrease in the rate of restenosis primarily by preventing the constrictive remodeling of the artery but at the cost of an increase in neointimal proliferation. The system used for intra-arterial beta-radiation therapy has Yttrium-90 beta ray emitting source (half-life, 64 hours; maximal energy, 2.284 Mev), a centering balloon and an automated delivery device. An 18-Gy dose not only prevents the renarrowing of the lumen typically observed after successful balloon angioplasty but actually induces luminal enlargement.

Drawback (all techniques)

- High cost of treatment (expensive equipment + time and labour-intensive)
- Lack of long term data (survival/late morbidity).

8. CyberKnife: **CyberKnife radiosurgery** is the noninvasive alternative to surgery for the precise treatment and effective removal of **cancerous tumours** (Fig. 47.5) from the body.

- The system has a miniaturised linear accelerator mounted on a robotic arm with 6 different points of axis where it can bend, turn, tilt or swivel with sub millimetre accuracy.
- Destroys tumours with highly precise beams of radiation, tumours virtually anywhere in the body, quickly, painlessly and without downtime or hospital stay in one to five sessions (hypofractionation).
- Thus offers new hope to patients who have inoperable or surgically complex tumours and for those who may be looking for an alternative to more invasive surgery.

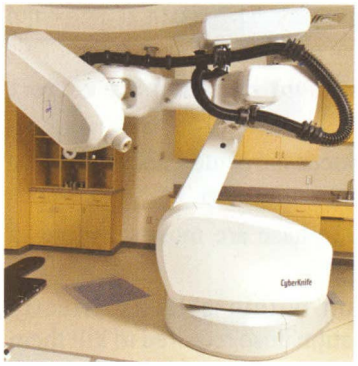


Fig. 47.5: CyberKnife

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated.
- New key boxes have been added.
- Recent advances in radiotherapy have been upgraded.

MULTIPLE CHOICE QUESTIONS

1. The following statements about radiation are correct *except*:

A. Fractional radiation reduces the acute effects of single doses of radiation on normal tissues

B. Cells in mitosis and G2 phase are most sensitive to radiation

C. Oxygen concentration within cells is directly proportional to the radiation dose required to kill them

D. Standard dose of radiation is 180–200 cGy per day and 5 per week over 4–6 cycles

2. A dose-response curve for radiation is of which shape?

A. Sigmoidal

B. Linear-rising

C. Bell-shaped

D. Irregular

3. The following are the principles of brachytherapy *except*:

A. It is a type of method of delivery of ionising radiation from sources inside or close to the tumour

B. External RT is used to deliver a higher dose (boost) to treat the primary site while brachytherapy treats the regional spread

C. It is highly localised, specific to a given tumour volume

D. It has a high dose rate, given as continuous RT in a single course

4. 1 cGy is:

A. 1 rad B. 10 rad

C. 100 rad D. 1000 rad

5. The most commonly used chemotherapeutic agent with radiation for head and neck cancers is:

A. Vincristine

B. Doxorubicin

C. Cisplatin

D. Etoposide

6. Which of the following statements regarding chemotherapy adjuvant to radiotherapy is false?

A. Agent of choice will have minimum toxic effects on the organs included in the radiation target volume

B. Neoadjuvant chemotherapy refers to 1–3 courses of CT after definitive RT

C. Adjuvant CT refers to a course after completion on definitive RT

D. Intrathecal CT is a prophylactic measure to prevent CNS metastasis in cancers like ALL or high grade lymphomas

7. IMRT refers to:

A. Intensity modified radiation therapy

B. Intensity modulated radiation therapy

C. Intensive method of radiation therapy

D. Intravascular method of radiation therapy

8. Intra-arterial brachytherapy uses which of the following?

A. Technetium-99

B. Yttrium-90

C. Iodine-123

D. Cobalt-60

9. The following is true about electron beam therapy *except*:

A. The source is mainly a linear accelerator

B. Most useful range is 6–20 Mev

C. Deeply situated tumours at a depth of > 5 cm are treated with this

D. Provides dose uniformity within the target volume

10. The following is true about postoperative RT:

A. Exact disease extent is known and treatment can be individualised

B. Postoperative complications, especially wound healing is affected

C. Operative margins are ill-defined

D. Potential for unnecessary radiation increases

ANSWERS

1 C 2 A 3 B 4 A 5 C 6 B 7 B 8 B 9 C 10 A

48

Principles of Anaesthesiology

- Preoperative assessment
- General anaesthetic agents
- Muscle relaxants
- Endotracheal intubation
- Monitoring
- Local anaesthetics
- Regional anaesthesia
- Pain relief
- Complications of anaesthesia
- Cardiopulmonary resuscitation
- What is new?/Recent advances

Surgery has been practised for ages. However, the advent of modern techniques of anaesthesia has allowed surgery to develop by leaps and bounds. If there is a well-informed, vigilant and safe anaesthesiologist taking care of the patient, the surgeon is able to concentrate on the surgical procedure unhindered. The anaesthetist of yore who was treated often as a technician assistant of the surgeon has metamorphosed into the present day anaesthesiologist, who is also a perioperative physician. An anaesthesiologist's understanding of anatomy, physiology and pharmacology, and his expertise of management of critical illness and pain have taken him into new fields such as Intensive Care, Pain and Palliative Care. Even then, the main realm of the anaesthesiologist remains the operation theatre.

A good understanding of the physiology and pharmacology, complemented by continuous and vigilant monitoring has made the practice of anaesthesia safer. Safe practice of anaesthesia comprises the following steps: a good rapport with the patient, a thorough preoperative preparation and premedication, monitoring and perioperative care.

PREOPERATIVE ASSESSMENT AND PREMEDICATION

Every patient is anxious when he comes into the operating room. The causes of anxiety can be varied: fear of the disease condition, surgery, anaesthetic and the anticipated pain. A good rapport developed between the patient and the anaesthesiologist can build up his confidence and help allay many of these fears.

Anaesthesia is associated with changes in the internal homeostasis. Normally these are well tolerated by the different systems. However, if the patient has a pre-existing derangement, his capacity to withstand changes in his internal milieu may be limited. It is thus very important to know the preoperative condition of the patient.

History

A detailed history of the patient with symptoms pertaining to the various systems must be elicited (Table 48.1).

Physical examination

A detailed physical examination is done and the relevant history specially borne in mind. General physical examination includes:

Vital signs: Blood pressure, heart rate, respiratory rate, temperature (and oxygen saturation, in relevant cases).

PICCLE: Pallor, icterus, cyanosis, clubbing, lymphadenopathy, oedema and raised jugular venous pulse.

Airway: Examine carefully to detect any difficult airway.

Spine: To rule out infection over the skin covering the spine, tenderness, stiffness or fractures of spine, to check spaces.

Veins: Ease of obtaining venous access is also assessed.

Systemic examination

A detailed examination of the various systems is then carried out and relevant findings noted.

Table 48.1 History taking prior to anaesthesia

Body system	History—Points to note
1. Cardiovascular system	Dyspnoea, angina, syncope, palpitations, pedal oedema, previously diagnosed to have a cardiac problem, effort tolerance
2. Respiratory system	Cough, fever, breathlessness, chest pain, recent onset upper respiratory tract infection
3. Central nervous system	Consciousness level, convulsions, orientation, ability to walk, speech, movement of all four limbs, bowel and bladder habits, any paraesthesia or altered sensation in the limbs.
4. Renal system	Decreased urine output, haematuria
5. Hepatic	Jaundice
6. Haematology	Easy bruising, increased blood loss with trivial injuries
7. Musculoskeletal	H/o weakness in the limbs
8. Previous medical history	Previous surgeries/ anaesthetics, any problems during the previous experience. H/o hospitalisation in the past, H/o tuberculosis, asthma, diabetes, hypertension, epilepsy, H/o current medications, steroid intake in the last 6 months
9. Allergy	Allergy to any drug or other substances
10. Addictions	Smoking, alcohol, drug abuse
11. Pregnancy	Last menstrual period/possibility of pregnancy (in female patients)
12. Family	Relevant family history, particularly pertaining to anaesthesia

Assessment of airway

Whenever a person becomes unconscious, the tongue and epiglottis fall back on to the pharynx and obstruct the airway. Since the anaesthesiologist makes the patient unconscious during a general anaesthetic, it is his duty to ensure that the patient's airway patency is maintained. Hence, assessment of the airway becomes important before a patient is made unconscious.

The assessment is done as follows:

The 1–2–3 test

- When a person opens his mouth, one should be able to insinuate one finger in the temporomandibular joint.
- There should be at least two finger breadths' distance between his incisors.
- There should be at least three finger breadths' distance between the chin and the thyroid cartilage (thyromental distance) of the patient.

Mallampati test

The patient is made to sit upright, open his mouth wide and protrude his tongue. The structures visualised are classified as given in Fig. 48.1. These classes roughly correlate with the following grades of laryngoscopic views (Fig. 48.2). Difficult airway may be anticipated in Mallampati Class III and IV and the anaesthesiologist must be prepared to secure airway in such patients.

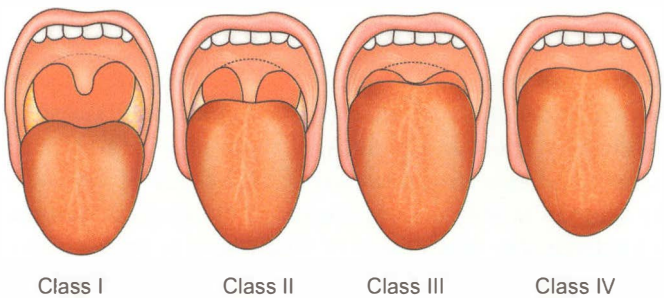


Fig. 48.1: Mallampati classification of the airway

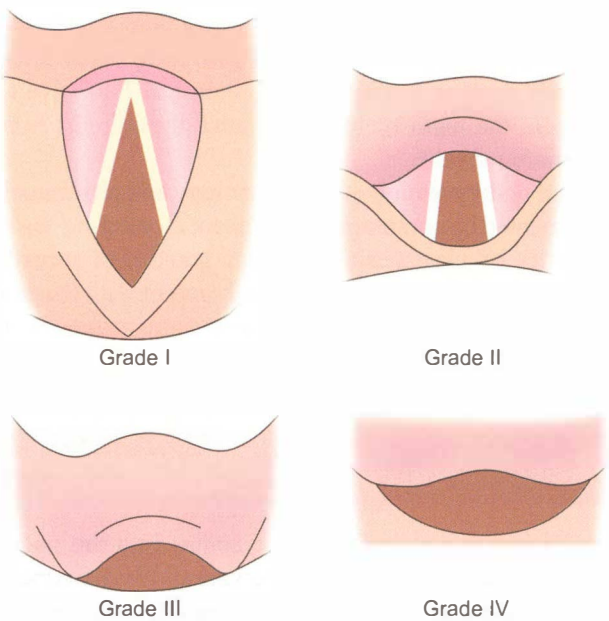


Fig. 48.2: Modified Cormack and Lehane grading of laryngoscopic view

Neck movements

Restriction of neck movements, especially extension makes endotracheal intubation more difficult.

Investigations

There is no place for routine investigations to be ordered before any surgery. Investigations that are required to assess the baseline status of the patient and have a bearing on the perioperative course must be done. Thus, they should be tailored to the individual patient. The common investigations ordered are as follows:

Haemoglobin estimation: All patients

Electrocardiogram, blood sugar estimation, blood urea, serum creatinine and electrolytes: Male patients > 40 years, smokers and female patients above 50 years.

Total and differential white cell count: If infection is suspected.

Platelet count and coagulation profile: If any abnormality is suspected or bleeding is anticipated.

Chest X-ray is requested for if a major abdominal or thoracic surgery is planned, postoperative ventilation is expected or cardiorespiratory disease is suspected.

Stress test, echocardiogram, pulmonary function test or a blood gas analysis are obtained as necessary.

Blood grouping and cross-matching are requested if the surgery may be associated with major blood loss.

ASA Physical Status Classification

The patient's preoperative physical status can be classified into five different categories. The American Society of Anaesthesiologists classification of Physical Status is as follows:

ASA I : Healthy patient, no medical problems

ASA II : Mild systemic disease

ASA III : Severe systemic disease, but not incapacitating

ASA IV : Severe systemic disease that is constant threat to life

ASA V : Moribund, not expected to survive without the operation

A suffix E is added if the surgery is of emergent nature. A sixth category called ASA VI is given to the patient who is declared brain dead whose organs are being removed for donor purposes.

The risks associated with anaesthesia in a patient with concurrent diseases must be assessed. Algorithms by different organisations are available to help decision-making in individual patients: For example, American Heart Association recommendations for evaluation of ischaemic heart disease in a patient posted for noncardiac surgery. The goal of preoperative assessment and preparation is thus to ensure that the patient is optimised and is in the best possible condition prior to surgery and anaesthesia.

Informed consent

The patient is explained about the planned anaesthetic, the problems anticipated and the risks involved in his own language and a **written and informed consent** is obtained.

Preoperative instructions

The patient is permitted to take solids and milk up to 6–8 hours and clear fluids up to 3 hours prior to surgery. This may be relaxed for neonates and infants where a four-hour fast for breast milk is sufficient. Formula feeds and milk from any other source are treated as solids. If the last food intake contained a considerable amount of fat, gastric emptying time may be delayed (Key Box 48.1).

KEY BOX 48.1

FASTING INSTRUCTIONS

• Adults and older children	Solids and milk: 6 hours Clear fluids: 3 hours
• Infants and neonates	Formula feeds: 6 hours Breast milk: 4 hours Clear fluids: 2 hours

Premedication

Premedication is the term given to the administration of certain medications prior to anaesthesia. The drugs used for premedication and their objectives are as follows:

1. **To allay anxiety:** Certain degree of anxiety is felt by most patients before surgery. A good rapport developed between the patient and the anaesthesiologist helps relieve this anxiety. Any of the following medications may be used to reduce anxiety.
   Adults: (Night before and morning of surgery)
   Tab Diazepam 0.1–0.2 mg/kg
   Tab Lorazepam 2–4 mg orally
   Children: To enable easy separation from parents.
   Midazolam 0.5 mg/kg (maximum 10 mg) mixed with 5 ml of paracetamol syrup is given orally 15–20 minutes prior to the procedure.
   Triclofos syrup, 100 mg/kg, one hour prior to surgery
2. **To relieve pain:** If the patient has any painful condition that could get aggravated on movement, a narcotic is often added to reduce the pain during shifting from the ward, e.g. fractures. Morphine (0.1 mg/kg), pethidine (0.5 mg/kg) or tramadol (0.5–1 mg/kg) given IM are often used.
3. **To dry secretions:** An anticholinergic such as **glycopyrrolate** (0.2 mg) is added if a fibreoptic intubation is planned so that oral secretions do not hinder vision. Local anaesthetic agents produce better local anaesthesia of the upper airway when the mucosa is dry. It may be given intravenously just prior to surgery in ENT surgeries and oral surgeries.

4. **To help anaesthesia induction:** Premedication with a narcotic provides analgesia and helps induce anaesthesia more smoothly.
5. **To blunt baroreceptor reflexes:** A small dose of β blockers or clonidine may be given in certain patients to blunt baroreceptor reflexes during intubation.
6. **To reduce gastric volume and acidity:** Some patients are at risk of regurgitation of gastric contents and aspiration. They may be premedicated with a prokinetic such as **metoclopramide** (10 mg) and a H_2 blocker such as **ranitidine** (150 mg orally). **Pantoprazole** 40 mg may be given instead of ranitidine. The consequences of aspiration of gastric contents depends on its quantity and acidity. Particulate matter, if aspirated can cause mechanical blockage of the airways. Metoclopramide reduces residual gastric content by hastening gastric emptying and ranitidine reduces the acidity. Sometimes, a nonparticulate antacid such as sodium citrate may be given to neutralise existing gastric acid. Multiple factors during anaesthesia and surgery increase the chances of nausea and vomiting. Since anaesthesia involves blunting of airway reflexes, patients are at risk of aspiration. This is why patients are kept fasting before elective surgery. This may not be possible for patients who are posted for emergency surgery. It is assumed that these patients are 'full-stomach' and appropriate precautions are taken.

TYPES OF ANAESTHESIA

Anaesthesia can be classified into two main categories: General anaesthesia and regional anaesthesia.

General anaesthesia (GA)

The patient is unconscious and there is a generalised and reversible depression of the central nervous system.

Regional anaesthesia

This involves injection of local anaesthetic agents in close proximity to the nerves or nerve bundles supplying the site of surgery. Regional anaesthesia can be central neuraxial block or peripheral nerve blocks.

Central neuraxial block

When regional anaesthesia is induced by injecting the local anaesthetic agents around the spinal cord, it is called central neuraxial block.

Peripheral nerve block

When regional anaesthesia is induced by injecting the local anaesthetic agents around nerve plexuses or individual nerves, it is called peripheral nerve block.

The choice of anaesthesia depends on several factors:

- The site and duration of surgery
- General condition of the patient
- Expertise of the anaesthesiologist and
- Preference of the patient.

Regional anaesthesia has a small failure rate and certain surgeries may outlast the duration of block provided by the regional block, in which case general anaesthesia may need to be given. Thus even though the primary anaesthetic contemplated is regional anaesthesia, patients must also be fit for general anaesthesia.

GENERAL ANAESTHETIC AGENTS

The term general anaesthesia includes:

- Hypnosis (sedation)
- Amnesia
- Analgesia and
- Muscle relaxation.

General anaesthetic agents are of two main types: Inhalational anaesthetic agents or intravenous anaesthetic agents.

INHALATIONAL ANAESTHETIC AGENTS

- **Volatile anaesthetics:** The volatile anaesthetic agents need a vaporiser to calibrate and deliver the vapour accurately in measured doses, e.g. halothane
- **Nonvolatile anaesthetics:** e.g. nitrous oxide

Classification

I. Agents of mainly historical interest

1. Ethyl chloride
2. Chloroform
3. Trichloroethylene
4. Cyclopropane
5. Methoxyflurane
6. Enflurane

II. Agents in occasional use

1. Diethyl ether

III. Agents in clinical use

1. Halothane
2. Isoflurane
3. Sevoflurane
4. Desflurane
5. Nitrous oxide

IV. Agent undergoing clinical trials—Xenon

A comparison of clinical effects of common inhalational anaesthetic agents in use is given in Table 48.2. Only important effects have been mentioned for the benefit of students.

HALOTHANE

This was a very popular anaesthetic agent till recently but is gradually being replaced by isoflurane. Reasons for dwindling popularity are:

- Myocardial depression
- Arrhythmogenicity
- Remote possibility of halothane hepatitis
- Easier availability of isoflurane (a safer agent).

Halothane hepatotoxicity

Two types of halothane induced hepatic dysfunction are recognised:

Type 1: It is mild, self-limiting and more common. It is associated with mild increases in liver enzymes but not jaundice. It is caused by reductive metabolism of halothane.

Type II: On extremely rare occasions (widely quoted incidence 1:35,000), the administration of halothane may be associated with the production of hepatitis. This entity, known as halothane hepatitis is largely a diagnosis of exclusion. It is fulminant with a high mortality rate (up to 50%). It is associated with fever, jaundice and grossly elevated liver enzymes. The toxic metabolite, trifluoroacetic acid reacts with liver proteins and triggers immune-mediated reaction in genetically susceptible individuals. Risk factors for development of halothane hepatitis are female gender, obesity, prior history of postanaesthetic jaundice, genetic susceptibility, repeated administration and enzyme-inducing drugs such as phenobarbitone.

Halothane is metabolised up to 20%, and sevoflurane (2%) and isoflurane (0.2%). Thus, hepatic dysfunction, although reported with sevoflurane and isoflurane, is extremely rare.

ISOFLURANE

- It is a halogenated ether.
- Isoflurane causes **less cardiac depression**. Hence, it is used in patients with cardiac disease.
- It causes **less cerebral vasodilatation** than halothane. Hence, it is preferred during neurosurgery.
- Isoflurane is now **routinely used** in all cases in many centres because it produces less arrhythmias and myocardial depression, is associated with only a remote risk of hepatitis and is not expensive.
- It has a **pungent** smell and hence cannot be used for inhalation induction.

SEVOFLURANE (Fig. 48.3)

- It is a newer general anaesthetic agent and has a sweet smell.
- Useful for **inhalation induction, especially in children.**
- Induction and recovery are faster than with halothane.
- It produces minimal myocardial depression.
- It is also a useful agent for induction of anaesthesia in patients with **difficult airways.**
- Its use is **limited only by its cost**.

DESFLURANE

- It is another new volatile anaesthetic agent.
- It causes vasodilatation and increase in heart rate.
- Induction and recovery are **very fast.**
- However, it is **irritant to the respiratory tract** and hence is not suitable for inhalational induction.
- It also requires a specially constructed **heated vaporiser** because of its high volatility.

Table 48.2 Comparison of different inhalation anaesthetics

Parameter	Halothane	Isoflurane	Sevoflurane
Smell	Pleasant	Pungent	Pleasant
Inhalation induction	Suitable	Not suitable	Suitable
Effect on CNS			
CNS depression	Rapid, progressive	Rapid, progressive	Rapid, progressive
Induction and recovery	Fast	Faster	Fastest
Effect on RS			
Minute ventilation	Depressed	Depressed	Depressed
Bronchodilatation	+++	++	++
Effect on CVS			
Myocardial contractility	↓↓↓	↓↓	↔
Heart rate	↓↓	↑↑	↔
Blood pressure	↓↓	↓↓↓	↓
Arrhythmias	+++	↔	↔
Effect on uterine muscle tone	↓↓	↓	↓
Effect on skeletal muscle	Relaxes	Relaxes	Relaxes

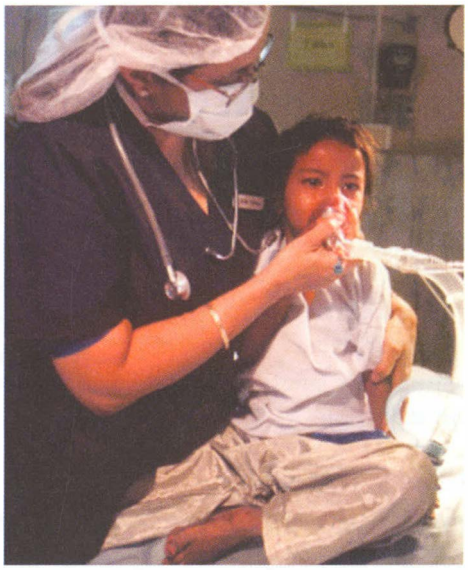


Fig. 48.3: Inhalation induction of anaesthesia in a child using sevoflurane. Note how the child is reassured by the comforting arm of the anaesthesiologist

NITROUS OXIDE

- It is an anaesthetic gas which is compressed and supplied as a **liquid in cylinders**.
- It is sweet smelling and nonirritant.
- It provides **analgesia** but is insufficient to produce an adequate depth of anaesthesia when used alone.
- It **enhances induction** of anaesthesia with the volatile anaesthetics and reduces their requirement.
- It does not produce significant depression of the cardiovascular system.

Uses

- It is used along with volatile anaesthetic agents as part of balanced anaesthesia.
- A combination of 50% nitrous oxide and 50% oxygen is available as Entonox. It is used to provide labour analgesia.
- It may also be used to provide analgesia for small procedures in dentistry.

Side-effects

- It can **diffuse into closed gas spaces** such as pneumothorax, obstructed intestines, sinuses and middle ear, and can cause barotrauma. The volume of a cavity can increase 3 to 4 times within 1–2 hours. Hence, nitrous oxide is best avoided in patients in whom such expansion may be anticipated.
- Its use is associated with increased incidence of **postoperative nausea and vomiting.**
- In prolonged administrations, it can affect vitamin B_{12} synthesis causing megaloblastic anaemia.
- **Teratogenicity:** This has been observed in pregnant rats exposed to nitrous oxide for prolonged periods but not proved in human beings. Nitrous oxide is best avoided in early pregnancy.

INTRAVENOUS ANAESTHETIC AGENTS

Intravenously administered anaesthetic agents are more popula for induction of anaesthesia because it is more rapid and smoot than that associated with inhalational agents. They can also b used for maintenance of anaesthesia, sedation during regiona anaesthesia, sedation in the ICU and treatment of statu epilepticus.

They can be classified into **rapidly-acting** (acting withi one arm-brain circulation time) and **slow-acting** (those tha take longer than one arm-brain circulation time) (Key Bo 48.2).

KEY BOX 48.2

INTRAVENOUS ANAESTHETICS

- Rapidly-acting: Thiopentone
  Propofol
  Etomidate
- Slower-acting: Ketamine
  High dose opioids
  Benzodiazepines

THIOPENTONE SODIUM

It is an ultra-short acting barbiturate, available as a yellowish powder. It is used as a 2.5% solution (25 mg/ml). It is used in a dose of 4–5 mg/kg intravenously.

Clinical effects

CNS

- Generalised depression of the CNS is observed within 15 to 20 s of IV injection of thiopentone. **Loss of eyelash reflex is used as an end-point.**
- It is a potent anticonvulsant.
- It is not an analgesic. To the contrary, it increases pain sensation **(antanalgesic)**. Consciousness is regained within 5 to 10 minutes.

CVS

- It produces myocardial depression, peripheral vaso-dilatation and hypotension especially when large doses are administered rapidly.
- Profound hypotension may occur, especially in a patient with hypovolaemia or cardiac disease. It may induce tachycardia.

RS

- It reduces respiratory drive. A short period of apnoea is common.
- It can precipitate asthma in susceptible individuals.

Skeletal muscle

There is poor muscle relaxation with thiopentone.

Uterus and placenta

- There is little effect on resting uterine tone.
- It crosses placenta rapidly, although foetal blood concentration is far less than that observed in the mother.

Eye

- It reduces intraocular pressure.
- The corneal, eyelash and eyelid reflexes are abolished.

Side effects

- Hypotension, tachycardia
- Respiratory depression
- Irritant to veins and can cause thrombophlebitis. If it extravasates, can cause tissue necrosis.
- Intra-arterial injection: Usually inadvertent, can cause severe arterial spasm and pain. This may be treated with vasodilators and heparin.
- Allergic reactions: Very rare.

Contraindications

Absolute	*Relative*
1. Airway obstruction	1. Hypovolaemia
2. Acute intermittent porphyria	2. Asthma
3. Previous hypersensitivity reaction	

PROPOFOL

This drug became commercially available in 1986. It is comparable to thiopentone but is five times more expensive. It is highly lipid soluble and is formulated in a white, aqueous emulsion containing soy bean oil and egg phosphatide. It is used in a dose of 2 to 2.5 mg/kg intravenously. The dose should be reduced in the elderly and in haemodynamically unstable patients.

Clinical effects

CNS

- Propofol depresses the central nervous system within 20 to 40 s of injection. **Loss of verbal contact is used as an end-point.**
- Recovery is rapid and there is a minimal "hang-over" effect even in the immediate post-anaesthetic period.

CVS

- The arterial pressure decreases more than with thiopentone. This is due to peripheral vasodilatation. The degree of hypotension can be reduced by slowing the rate of administration.
- Heart rate increases slightly.

RS

- After induction, apnoea occurs commonly and for a longer duration than with thiopentone.
- It causes ventilatory depression, particularly with opioids.
- It has no effect on bronchial muscle tone.

GIT, uterus and placenta: Propofol has no effect on GI motility but causes mild transient decrease in hepatorenal function.

Uses (Table 48.3)

- As an **induction agent**
- As an infusion, can be used to provide **total intravenous anaesthesia**
- To provide **conscious sedation**
- As continuous infusion to **sedate patients in the ICU**, since its half-life is very short.

Adverse effects

- Cardiovascular depression
- Respiratory depression
- Pain on injection
- Allergic reactions.

ETOMIDATE

- Etomidate is a rapidly acting intravenous anaesthetic agent with a short duration of action of 3–5 minutes.
- It is a **very cardiostable** agent and is used to induce anaesthesia in cardiac patients and in haemodynamically unstable patients.
- However, continuous infusions are not advisable as it is known to depress synthesis of cortisol by the adrenal gland and impair response to ACTH.
- It is used in a dose of 0.2–0.3 mg/kg IV.

Adverse effects

1. **Suppression of synthesis of cortisol with infusions.**
2. Excitatory phenomena: Involuntary movements, cough
3. Pain on injection and venous thrombosis
4. Nausea and vomiting

KETAMINE HYDROCHLORIDE

Ketamine differs from other intravenous anaesthetic agents in many respects and produces dissociative anaesthesia, rather than generalised depression of the central nervous system. It is used in a dose of 1–2 mg/kg IV or 4–5 mg/kg IM.

Clinical effects

CNS

- It induces anaesthesia within 30–60 s of intravenous injection and lasts for 10–20 mins. It is effective within 3–4 mins of intramuscular injection and lasts for 15–25 mins.

Table 48.3 Comparison of different intravenous anaesthetics

Parameter	Thiopentone	Propofol	Ketamine
Onset	Rapid	Rapid	Slow
Effect on CNS			
CNS depression	↓↓↓	↓↓↓	Dissociative anaesthesia
Cerebral blood flow	↓↓	↓↓	↑↑
Pain sensation	Antanalgesia	No change	Profound analgesia
Hallucinations	None	None	Present
Effect on RS			
Minute ventilation	↓	↓↓	↔
Bronchodilatation	Can precipitate bronchospasm	No effect	Bronchodilator
Airway reflexes	Not blunted	Blunted	Maintained
Effect on CVS			
Myocardial contractility	↓	↓	↑↑
Heart rate	↑	↑	↑↑
Blood pressure	↓	↓↓	↑↑
Arrhythmias	↔	↔	↔
Muscle tone	↓	↓	↑
Intraocular pressure	↓	↓	↑

- It produces anaesthesia by dissociating the cerebral cortex from the limbic system.
- It is a potent analgesic.
- It increases cerebral blood flow and intracranial pressure.

CVS
The heart rate, blood pressure and cardiac output increase.

RS
- Respiration is usually well-maintained with ketamine although transient apnoea may occur occasionally.
- Ketamine is a good **bronchodilator**.

Skeletal muscle
There is increased muscle tone. Some spontaneous and involuntary movements may occur.

GIT
Increased salivation can occur and can be prevented by using anticholinergic agents.

Eye
Intraocular pressure increases.

Uterus and placenta
It readily crosses the placental barrier and hence should be given in lower doses in pregnant patients.

Adverse effects
- Emergence delirium
- Hypertension and tachycardia
- Increased intracranial pressure
- Vivid and unpleasant hallucinations are known with ketamine and can be prevented by prior injections of benzodiazepines.

Uses (Table 48.3)
- Patients in severe hypotension, shock
- Paediatric anaesthesia
- Analgesia and sedation
- Bronchial asthma
- **Difficult locations:** Accident sites, war casualties.

PHYSIOLOGY OF NEUROMUSCULAR JUNCTION

When a nerve impulse arrives at the neuromuscular junction through the motor neuron, acetylcholine (ACh) molecules are liberated from the nerve endings into the junctional cleft. Acetylcholine molecules act as neurotransmitters by interacting with the **nicotinic ACh receptors** on the postjunctional muscle membrane at the motor end plates. This induces the opening of ionic channel of the receptor to allow ionic (sodium, calcium) flux into the muscle. The sudden influx of sodium results in depolarisation and muscle contraction.

Acetylcholine receptor
The acetylcholine receptor is flower-shaped with five petal-like structures. These five subunits are named $\alpha(2)$, $\beta(1)$, $\varepsilon(1)$ and $\delta(1)$. Each alpha subunit must be occupied by an acetylcholine molecule to open the channel. This acetylcholine

receptor at the neuromuscular junction is nicotinic in nature and thus different from those in the rest of the body.

MUSCLE RELAXANTS

These are drugs that interfere with the combination of acetylcholine molecules with their receptors. These block neuromuscular transmission and cause relaxation of the muscle resulting in muscle paralysis.

Neuromuscular blockers are of two types: **Depolarising** and **nondepolarising muscle relaxants**.

DEPOLARISING MUSCLE RELAXANTS (SUCCINYL CHOLINE)

It is the only depolarising muscle relaxant in clinical use. It has a molecular structure similar to acetyl choline. It combines with the alpha subunit of the ACh receptor and produces muscle contraction. However, unlike acetyl choline, it has a prolonged action. Continued depolarisation of a muscle results in **accommodation blockade** and the muscle relaxes.

Dose: 1–1.5 mg/kg intravenously.

Onset of action: Within 60 seconds

Duration of action: 3–5 minutes

Metabolism: By plasma cholinesterase.

Uses

Succinyl choline is the only muscle relaxant which has the **shortest time to onset of action** (60 seconds) **and shortest duration of action** (3–5 minutes). It is used:

- To facilitate endotracheal intubation in 'full-stomach' patients.
- It is useful in patients with difficult airway because it gives very good relaxation to facilitate intubation but if intubation fails, the patient is likely to resume spontaneous breathing early and hypoxic brain injury may be avoided.
- To maintain paralysis for short procedures.

Adverse effects

- **Muscle pains:** The initial depolarisation that occurs due to succinyl choline causes uncoordinated contraction (fasciculations) of different groups of muscle fibres. This can cause severe muscle pain postoperatively.
- **Bradycardia,** especially if a second dose of succinyl choline is given. It is easily avoided by pretreatment with atropine.
- **Hyperkalaemia** in patients with renal failure, burns, massive crush injury, etc.
- **Increase in intracranial pressure**
- **Increase in intraocular pressure**
- **Prolonged action** in patients deficient in pseudocholinesterase (plasma cholinesterase). This occurs as a genetic problem in a small number of patients but may be an acquired problem as in severe liver disease.
- **Malignant hyperthermia** is a disorder that is unique and life-threatening disorder precipitated by exposure to **inhalation anaesthetics and succinyl choline** in susceptible patients. In malignant hyperthermia, the metabolic rate of muscle cells is increased tremendously due to a defective **ryanodine** receptor which is necessary for **reuptake of calcium** after a depolarisation.
- Those with a family history of anaesthetic mishaps, neuromuscular diseases such as muscular dystrophy may be susceptible to this disorder and succinyl choline is best avoided in these patients. It is not always possible to identify latent muscular dystrophies in infants and children. Administration of succinyl choline in these patients can be disastrous. Hence, the use of succinyl choline should be avoided in children less than 2 years unless an indication such as a full stomach exists. Even then, rocuronium may be considered as an alternative.

NONDEPOLARISING MUSCLE RELAXANTS

Pancuronium, vecuronium, rocuronium, atracurium and cisatracurium are drugs belonging to this group in clinical use. The nondepolarising muscle relaxants combine with the ACh receptors but do not have any intrinsic effect on the muscle. They cause muscle paralysis by preventing the acetyl choline molecules that are released from the nerve terminal from combining with the acetyl choline receptors on the postsynaptic membrane and producing their action (competitive inhibition). Even if one alpha subunit is combined with a molecule of nondepolarising muscle relaxant, the muscle cannot contract in response to a nerve impulse and gets paralysed. The muscle regains its power when the muscle relaxant gets metabolised.

The nondepolarising muscle relaxants can be classified according to their duration of action as follows:

- **Short-acting (10–20 min):** Mivacurium
- **Intermediate-acting (20–30 min):** Atracurium, vecuronium and Rocuronium
- **Long-acting (> 45 min):** d-Tubocurarine and pancuronium.

Uses

1. To facilitate endotracheal intubation
2. To maintain paralysis during anaesthesia and in the ICU.

REVERSAL OF NEUROMUSCULAR BLOCKADE

At the end of anaesthesia, the muscle relaxation produced by the nondepolarising muscle relaxant is usually reversed. This is to ensure good recovery of muscle power to maintain airway and respiration.

Acetylcholine molecules are broken down by cholinesterases. Anticholinesterases such as neostigmine block the action of cholinesterase, thus allowing acetyl choline molecules to accumulate in the neuromuscular junction. The block produced by the nondepolarising muscle relaxants is competitive. When acetyl choline molecules increase in number due to the action of the anticholinesterase, and nondepolarising muscle relaxant molecules decrease in number due to metabolism, muscle power returns. **Neostigmine** is the only anticholinesterase in clinical use. Its dose is 0.05 mg/kg body weight. Neostigmine increases the amount of acetyl choline not only at the neuromuscular junction but also in the entire body. It can cause the muscarinic effects of acetyl choline such as bradycardia, bronchoconstriction, etc. Hence, neostigmine is always combined with atropine (0.025 mg/kg) or glycopyrrolate (0.01 mg/kg) to counter the muscarinic effects.

Adequate recovery from neuromuscular blockade

Clinical indicators

- Opening of eyes without furrowing of forehead
- Good hand grip
- Raising arms against gravity
- Good cough
- Ability to lift head against gravity (sustained head-lift) for at least five seconds.

Objective criteria

The amount of neuromuscular blockade can be checked using a peripheral nerve stimulator. The ulnar nerve is stimulated and the response of the adductor pollicis muscle is checked. If good muscle contractions are seen in response to the nerve stimulation, the muscle power is said to have returned. Similarly, posterior tibial nerve and facial nerve may also be used to monitor neuromuscular junction.

The patient is allowed to breathe spontaneously and his trachea extubated only when there is clinical evidence of complete recovery from neuromuscular blockade. If there is inadequate recovery of muscle power, the patient may need to be ventilated artificially till muscle power is normal.

AIRWAY MANAGEMENT

General anaesthesia involves induction of unconsciousness during which the patient's ability to maintain his airway and breathing are impaired. It thus becomes necessary that the anaesthesiologist maintains and protects the patient's airway. In addition, not only anaesthesia but also oxygenation and ventilation are maintained well only if the airway is patent and secured.

Management of airway is one of the basic skills acquired by an anaesthesiologist. This is relatively easy in most individuals. Difficult airway is the term given when there is difficulty with mask ventilation, endotracheal intubation, or both.

BAG-MASK VENTILATION

When a person becomes unconscious, the tongue and the epiglottis fall back and obstruct the airway. Since they are attached to the mandible, lifting up of the mandible also lifts these two structures and opens airway. This is usually done by head-tilt and chin-lift method as during cardiopulmonary resuscitation. Jaw thrust is more popular with anaesthesiologists. The patient may be ventilated using a 'bag' (either a self-inflating bag or anaesthetic circuit) and a facemask (Fig. 48.4). The mask is triangular in shape, the narrow portion of which is placed at the bridge of the nose and the base placed in the depression between the lower lip and chin. The mask is held in place by an E-C technique (Fig. 48.5).

ENDOTRACHEAL INTUBATION

Endotracheal intubation is the most definitive way of maintaining airway in patients who require muscle paralysis and require intermittent positive pressure ventilation. It involves introduction of a tube into the trachea for maintaining the patency and protecting the airway as well as to ensure adequate oxygenation and ventilation. Whenever general anaesthesia is induced and needs to be maintained for long periods, endotracheal intubation is done.

Indications

- To administer general anaesthesia for long (>1–2 h) periods.
- To maintain patency of the airway in unconscious patients.

Fig. 48.4: Maintenance of airway and ventilation using bag (anaesthetic circuit) and mask

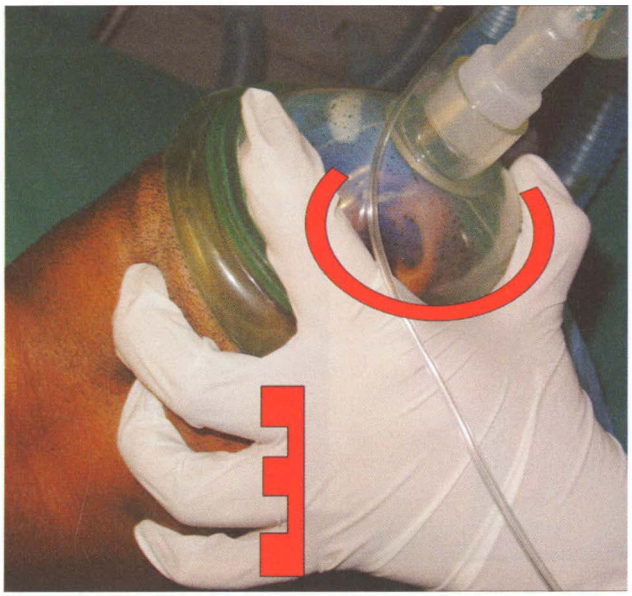


Fig. 48.5: The E-C technique of holding a face mask to get airway seal. The thumb and index fingers form a 'C' at the top of the mask holding it firmly down on the face. The middle, and ring fingers lift up the mandible as well as provide head tilt. The little finger aids in providing jaw thrust. Note that the finger pressure should be given on the bony part and not on the soft tissues under the chin

- To protect lungs from aspiration of regurgitated gastric contents.
- To ensure delivery of adequate tidal volumes to the lungs.
- To clear excessive and retained secretions from the lungs.

Contraindications

Endotracheal intubation may be extremely difficult and even dangerous. A tracheostomy may be better in such situations.

- When the upper airway integrity is lost as in extensive maxillofacial injury with bilateral fractures of mandible and maxillae.
- Injuries to the neck with laryngeal rupture
- Large tumours of the upper airway.

Equipment

Laryngoscopes

These consist of a handle and a blade.

- The handle contains batteries. The blade has a flange to push the tongue towards the left side. This ensures more room for visualisation of the glottis. A bulb nearer the tip of the blade lights up when the handle and blade are at right angles to each other and electrical contact is made.
- There are several types of laryngoscopes to aid in different situations.
  - **Macintosh type blade** (Fig. 48.6) **is curved** and is popular for use in adults.

Fig. 48.6: Macintosh laryngoscope. Note the curved blade. Sizes 1, 2 and 3 are displayed

Fig. 48.7: McCoy laryngoscope. Note the retractable tip of the laryngoscope blade

  - **The Miller blade is straight** and is used in children and in adults with difficult airway.
  - **The McCoy laryngoscope** (Fig. 48.7) has a **tiltable tip**.
  - **Bullard laryngoscope** for use in patients with cervical spine immobility.
  - **Fibreoptic laryngoscopes are flexible.**
  - **Videolaryngoscopes** have a small camera on the blade a little proximal to the tip and provide a better view of 'difficult to see' larynx.
  - **Short-handled laryngoscopes** are available for use in difficult airways, e.g. pregnant women, obese patients.

Fig. 48.8: Endotracheal tube. (A) Standard 15 mm connector (B) Pilot balloon (C) Inflatable cuff (D) Beveled patient-end (E) Black line to indicate optimum positioning of the tube. This line is usually placed just above the glottis

Endotracheal tubes (Fig. 48.8)

It is a 'C'-shaped tube and is commonly made of polyvinyl chloride (PVC). The machine end has a standard 15 mm diameter connector. The patient end is bevelled and has an opening on the side just proximal to the tip called the Murphy's eye. This ensures patency of the tube even if the bevelled tip is against the tracheal wall.

KEY BOX 48.3

ENDOTRACHEAL TUBE SIZE

• Adult male	8 or 8.5 mm/D
• Adult female	7 or 7.5 mm/D
• Children < 6 years	Age/3 + 3.5 mm/D
• Children > 6 years	Age/4 + 4.5 mm/D

Endotracheal tube size

Endotracheal tubes are available in different sizes. Their size is indicated as the internal diameter in mm (Key Box 48.3). However, the correct size will depend on the growth of the child.

Depth of insertion

- The distance of several points on the tube from the patient end is marked in cm along the tube. The tube is fixed at 22 or 23 cm in adult men and at 20 or 21 cm in adult women.
- In children, the following formula is used:
  Age/2 + 12 cm.

The tube should be positioned such that its tip must lie above the carina but well below the glottis.

Position

A pillow (7–10 cm) under the patient's head enables mild flexion at the cervical spine. The head is then extended at the atlanto-occipital joint. This is called the intubating position or "sniffing position".

Fig. 48.9: Oral endotracheal intubation

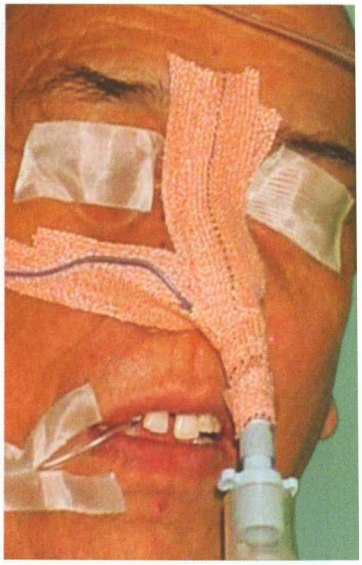


Fig. 48.10: Nasal endotracheal intubation

Route

Endotracheal intubation can be done either orally or nasally (Figs 48.9 and 48.10). It can be done either under direct vision or indirectly using a fibreoptic scope. It may need to be done blindly when visualisation of the glottis by direct means is not possible and a fibreoptic scope is not available.

In such cases, if the regular antegrade technique (mouth or nose to larynx) is not possible, retrograde intubation (larynx to mouth) may be tried. In the retrograde technique, a guide wire is passed from the cricothyroid membrane upward into the mouth or nose and the endotracheal tube is guided over it.

Procedure (Key Box 48.4)

The patient's head is placed in the sniffing position. The mouth is opened and the laryngoscope blade is introduced through the right angle of mouth along the tongue into the pharynx. Once the epiglottis is seen, the tip of the laryngoscope blade is pressed into the vallecula. This lifts up the epiglottis to reveal the glottis. The glottis is identified by the two pearly white vocal cords. Once the cords are seen, the endotracheal tube is inserted between them into the trachea.

It may also be done with the patient awake after administering local anaesthesia to the upper airway when a difficult intubation is anticipated.

KEY BOX 48.4

ENDOTRACHEAL INTUBATION

- Orotracheal or nasotracheal
- Direct or indirect (using fibreoptic laryngoscope)
- Under vision or blind
- Anaesthetised or awake
- Antegrade or retrograde

Many airway adjuncts are available for use when a difficult airway is encountered, especially when it is unanticipated. These include oropharyngeal airway, nasopharyngeal airway, laryngeal mask airway and Combitube.®

Confirmation of correct placement of endotracheal tube

Correct position of the endotracheal tube may be confirmed by the following

- Endotracheal intubation under vision
- Bilateral visible chest rise
- Bilateral equal air entry in the lungs
- Absent breath sounds in the epigastrium
- A square wave normal capnogram—gold standard
- Prompt inflation of the deflated bulb of oesophageal detector device.

Complications

The complications of endotracheal intubation may be classified as follows

Immediate

- Trauma to teeth, lips, tongue, pharynx or larynx
- Haemodynamic changes—tachycardia, hypertension, myocardial ischaemia
- Misplaced tube—accidental extubation, oesophageal intubation

Delayed

Laryngeal granuloma, laryngeal or subglottic stenosis

Oropharyngeal airway (Figs 48.11 and 48.12)

- It is available in various sizes. The correct size is chosen such that when it is placed along the side of the patient's face, it should extend from **the angle of mouth up to the tragus**. This is inserted along the tongue and reaches up to the posterior pharyngeal wall.
- Care should be taken not to push the tongue backwards with the airway itself.
- To avoid that, the airway is inserted with its concavity towards the palate and then turned once 50% of it is inserted and passed further.
- Please note that an oropharyngeal airway is excellent in patients who are completely unconscious. Patients who are conscious do not tolerate it. In patients who are semiconscious, it may initiate a gag reflex and induce vomiting. In such patients, a nasopharyngeal airway is a better choice.

Nasopharyngeal airway (Figs 48.13 and 48.14)

- It is a soft tube made of latex or silicon. It is available in various sizes. The correct size is the same size as an endotracheal tube for that patient.
- The correct length is chosen such that when it is placed along the side of the patient's face, it should extend from

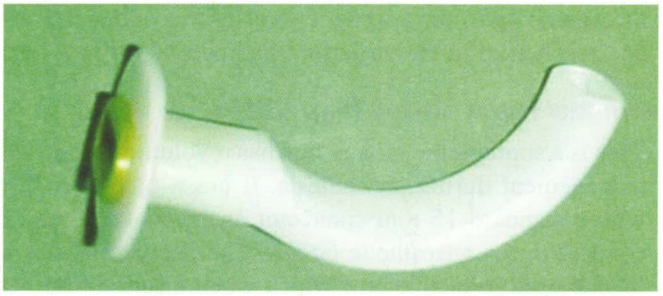


Fig. 48.11: Oropharyngeal airway

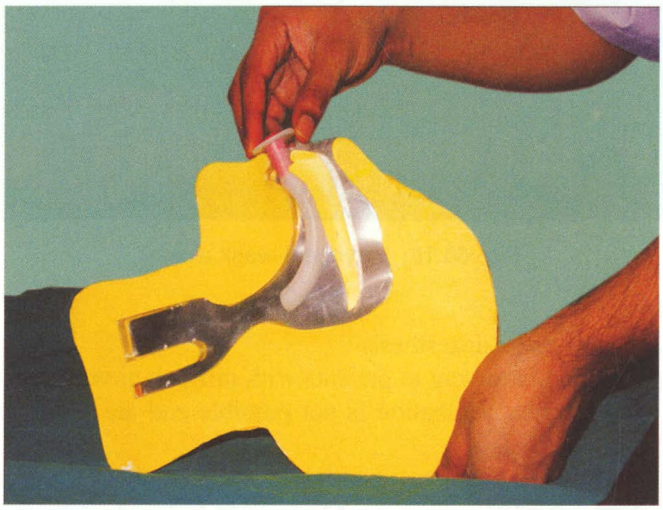


Fig. 48.12: Insertion of oropharyngeal airway. Take care not to push the tongue backwards

Fig. 48.13: Nasopharyngeal airway

Fig. 48.14: Insertion of nasopharyngeal airway

the **nostril up to the tragus**. This is inserted along the nose and reaches up to the posterior pharyngeal wall.

Laryngeal mask airway (Fig. 48.15)

- This is a supraglottic airway that has revolutionised airway management during anaesthesia. It has a tube or a shaft with a standard 15 mm connector at the proximal end to connect to the anaesthetic circuit. The patient end has an oval-shaped inflatable cuff which when inserted rests just above the larynx and creates an airtight seal.

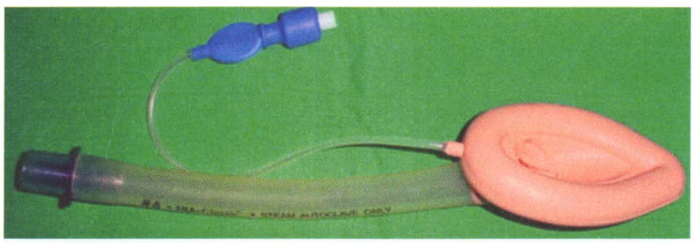


Fig. 48.15: Laryngeal mask airway

Uses

- To administer anaesthesia
- To maintain airway in patients with difficult airway where endotracheal intubation is not possible and are at risk of hypoxia
- It may also be used as a conduit for passage of endotracheal tube, either blindly or with the use of fibreoptic bronchoscope.

Endotracheal extubation

Endotracheal extubation is as important as intubation. A patient's trachea may be extubated when the following criteria are met

- Oxygenation and ventilation are satisfactory
- Patient is haemodynamically stable.
- Patient is fully conscious
- Able to maintain his airway patency
- Airway reflexes are intact
- Able to cough and clear airway.

Equipment for reintubation and personnel skilled in intubation should be readily available. After a good oropharyngeal suction to clear the secretions, the cuff is deflated and the tube removed. Oxygen should be administered by face mask and the patient monitored till he is stable and ready to go to the ward.

MONITORING IN ANAESTHESIA

The administration of anaesthesia is associated with changes in the internal homeostasis of the patient, especially the cardiac and respiratory systems. Constant monitoring of the various body systems is necessary to ensure the well-being of the patient and prompt recovery from anaesthesia at the end of surgery. Identification and prompt treatment of any untoward changes should alleviate complications due to anaesthesia.

- Monitoring used in anaesthesia may be classified into: **noninvasive and invasive monitoring.** The extent of monitoring depends on the patient's preoperative condition, the extent of surgery, the type of anaesthesia and the facilities available.
- Most patients are noninvasively monitored. Invasive monitoring becomes necessary when considerable haemodynamic instability exists or is expected to occur perioperatively.
- **Minimum monitoring standards** (according to guidelines from Indian Society of Anaesthesiologists) for a patient undergoing any type of anaesthesia are:
  - Noninvasive blood pressure (NIBP)
  - Electrocardiogram (ECG)
  - Pulse oximetry
- Monitoring capnography although not mandatory in India, is an important monitor in patients undergoing anaesthesia. In addition, monitoring of body temperature, anaesthetic gases and neuromuscular blockade is desirable.

NONINVASIVE MONITORING

Basic noninvasive monitoring includes **clinical observation of the patient** and monitoring of the patient's haemodynamic parameters. **Adequate cardiac output is associated with good urine output, warm and well-perfused peripheries and good capillary refill.** Heart rate may be monitored with a finger on the pulse. Blood pressure may be measured using a standard sphygmomanometer. More conveniently and accurately, heart rate and rhythm are continuously monitored using the electrocardiogram and blood pressure using automated noninvasive blood pressure monitoring systems.

Baseline readings of the heart rate, blood pressure and oxygen saturation prior to induction of anaesthesia are recorded. After induction, the patients are continuously monitored and a record of these is made at least every five minutes thereafter. When a patient undergoes an extensive or a long procedure associated with large fluid shifts, his urinary bladder is catheterised so that hourly urine output can be measured. A normal person produces at least 0.5–1 ml/kg/hour of urine output.

ELECTROCARDIOGRAM (ECG)

The objectives of electrocardiographic monitoring (Fig. 48.16) are to monitor heart rate and rhythm and to look for myocardial ischaemia. Lead II is monitored commonly as it is best for identifying arrhythmias. V5 detects left ventricular ischaemia. Hence, both Lead II and V5 should be monitored continuously for patients suspected to have ischaemic heart disease. Subtle changes in the electrocardiogram on the monitor must be confirmed by a 12 lead—ECG.

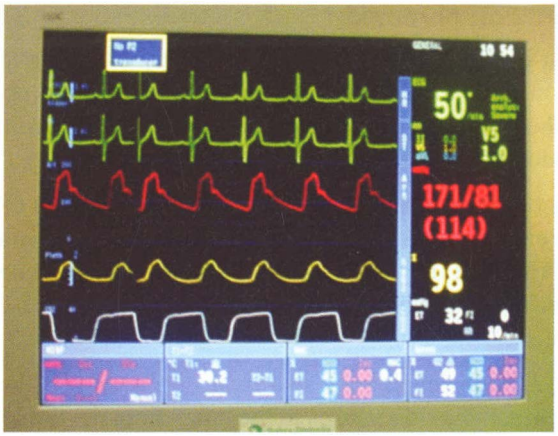


Fig. 48.16: Multimodular monitor

NONINVASIVE BLOOD PRESSURE (NIBP)

Most automated devices use an oscillotonometric technique and as a result the most accurate pressure is the mean arterial pressure. They tend to overestimate at low pressures and underestimate at high pressures. They may also give erroneous results in patients with atrial fibrillation or with other arrhythmias. The cuff width (Fig. 48.17) is the most important determinant of the accuracy of the pressure reading. The cuff width should be 40% of the mid-circumference of limb (the length should be twice the width). Cuffs which are too narrow tend to overestimate BP while those which are too wide tend to underestimate BP.

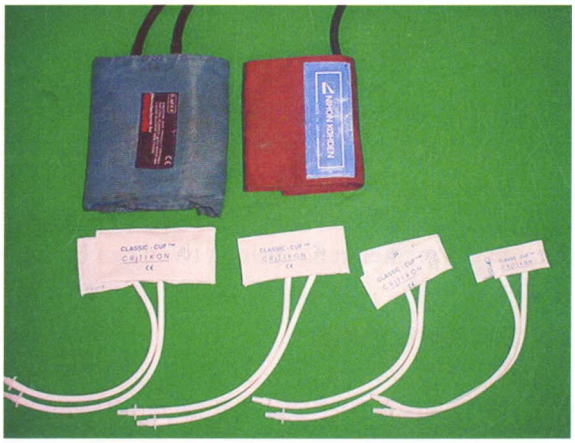


Fig. 48.17: Blood pressure cuffs of various sizes

PULSE OXIMETRY

The pulse oximeter displays the oxygen saturation of haemoglobin continuously, noninvasively and in real-time (Figs 48.18 and 48.19). Its use has revolutionised the way patients are monitored, both perioperatively and in the ICUs.

Working principle

The pulse oximeter works on the principle of **Beer-Lambert's law**. *This states that the amount of light absorbed by a solution is directly proportional to the amount of solute in it.* The pulse oximeter consists of two light emitting diodes (LEDs), emitting light at around **660 nm** and **940 nm**. Oxyhaemoglobin (OxyHb) absorbs more of light at 940 nm whereas reduced haemoglobin (Hb) absorbs more light at 660 nm. The amount of OxyHb in tissues increases with each pulse wave.

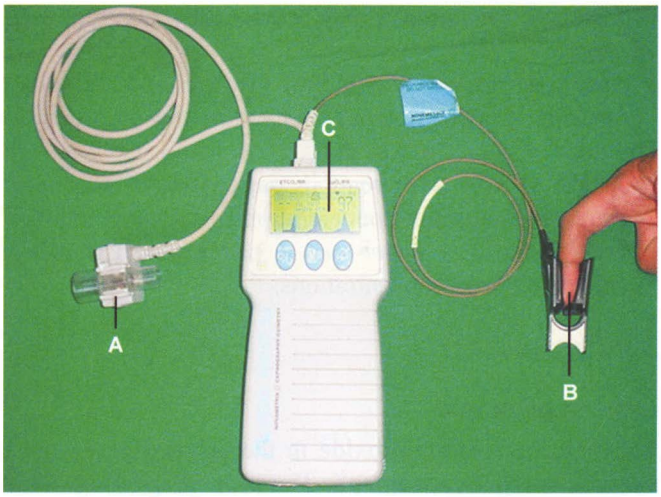


Fig. 48.18: A portable pulse oximeter and capnograph. (A) The capnograph and adaptor, (B) Pulse oximeter probe, (C) Graphical display window that can be programmed to show either a plethysmograph (as seen in the photograph) or a capnogram

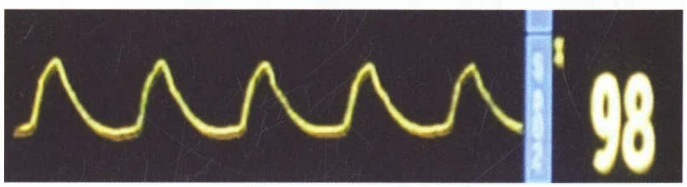


Fig. 48.19: The plethysmograph and oxygen saturation (SpO_2) obtained by a pulse oximeter

The differential absorption of light by OxyHb and reduced Hb and the varying concentration of these two substances with arterial pulsations are utilised in the construction of the pulse oximeter.

The pulse oximeter has one LED on, then the other on, then both off. This happens several times a second. The ratio of absorption of these two wavelengths of light by OxyHb and reduced Hb is related to the oxygen saturation using a nomogram.

Significance

- The normal oxygen saturation of a person breathing room air is around 98–100%.
- If the saturation dips below 90% or lower, hypoxia is said to be imminent. Saturation of 90% corresponds to PaO_2 of 60 mmHg.
- Hence, the inspired oxygen concentration is adjusted to maintain a SpO_2 of > 95% during anaesthesia.

Sites of monitoring

Fingers, toes, ear lobes; soles and palms in neonates/infants.

Limitations

- **Movement:** The pulse oximeter may not be reliable when there is movement of the part monitored.
- **Electrical interference** may occur with concurrent use of diathermy.
- **Vasoconstriction:** It may fail to pick up a pulse with profound vasoconstriction as in hypothermia or low cardiac output.
- **COHb:** It may over-read in patients with significant carboxyhaemoglobin concentrations.
- **Meth-Hb:** It may under-read in patients with significant methaemoglobin concentrations.

CAPNOGRAPHY

Working principle

The amount of carbon dioxide in the exhaled gases can be measured using a capnograph. The capnograph (Fig. 48.20) passes infrared light through the gases to be analysed. The carbon dioxide in the gases absorbs the infrared light in proportion to its concentration. This absorption is measured and the carbon dioxide concentration is displayed according to a nomogram. When the carbon dioxide (CO_2) concentration is displayed against time, a capnogram is obtained.

The normal capnogram

- The normal capnogram has four different phases as illustrated in Fig. 48.20.
- **Phase I:** Dead space gases
- **Phase II:** Mixed alveolar and dead space gases
- **Phase III:** Alveolar gases
- **Phase IV:** Inspiration
- The CO_2 concentration at the end of expiration is called end-tidal CO_2 concentration (**normal range: 35–40 mmHg**) which represents the alveolar carbon dioxide concentration. Arterial CO_2 concentration is 0–5 mmHg more than the end-tidal CO_2 concentration.

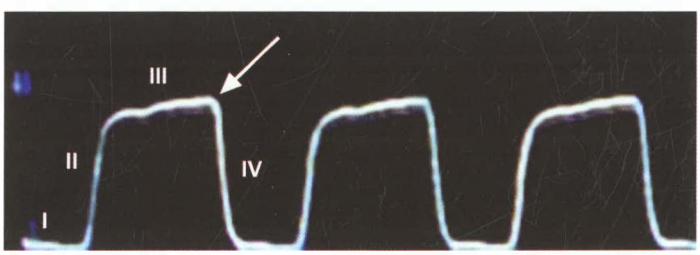


Fig. 48.20: The normal capnogram: Phase I—dead space gases, Phase II—mixed dead space and alveolar gases, Phase III—alveolar gases, Phase IV—inspiration. The arrow is pointing towards the end-tidal carbon dioxide concentration ($ETCO_2$)

Information obtained

Carbon dioxide is excreted from the body only through one organ in the body, i.e. lungs. A normal capnogram obtained from the exhaled gases helps identify the correct position of the endotracheal tube.

- If the capnograph registers **zero** levels of carbon dioxide, the endotracheal tube is malpositioned as in **oesophageal intubation or accidental extubation.** Mechanical **ventilator disconnection** also results in zero $ETCO_2$ levels.
- The $ETCO_2$ levels may also reduce to **near zero** levels if no carbon dioxide returns to the lungs from the tissues as in **profound hypotension and an impending cardiac arrest** may be suspected.
- However, **the $ETCO_2$ will register zero if the patient is in cardiac arrest** since there is no circulation and no carbon dioxide is brought to the lungs. In such situations, a capnograph cannot be used to identify the correct position of endotracheal tube. The capnograph will register CO_2 only if effective CPR is being given.
- However, a capnograph is extremely useful in assessing the effectiveness of CPR. **If $ETCO_2$ is > 15 mmHg during CPR, chances of successful resuscitation are higher.**
- A capnograph is also useful to detect hypo or hyperventilation, bronchospasm and recovery from neuromuscular blockade.

Limitations

- When there is an increase in physiological dead space ventilation (that part of ventilation that does not take part in gas exchange), the arterial to $ETCO_2$ gradient increases.
- In such situations, arterial blood gas analysis can be done to determine the gradient and then its trend can be followed.

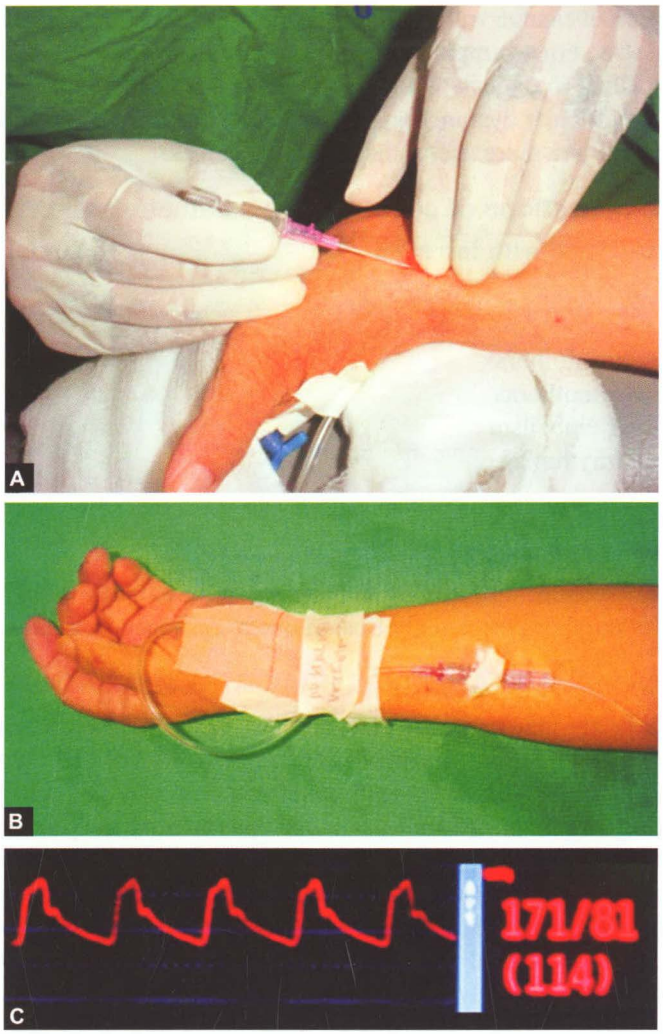
INVASIVE MONITORING

DIRECT ARTERIAL PRESSURE MONITORING

Direct arterial pressure monitoring is preferred whenever haemodynamic instability exists or is expected to occur perioperatively. In such situations, invasive measurement enables beat to beat monitoring of arterial pressure. A cannula is inserted into an artery and connected to a pressure transducer. The monitor displays beat to beat blood pressure both numerically as well as graphically.

Sites

- **Radial artery:** Most common (Fig. 48.21)
- **Dorsalis pedis artery:** When radial arteries are inaccessible or they have already been used.



Figs 48.21A to C: Radial arterial cannulation: (A) Cannulation, (B) Catheter securely taped in place and labelled 'Arterial line—Do Not Inject!', (C) The arterial waveform and pressure

- **Femoral artery**: If the peripheral circulation is sluggish, blood pressures are too low and the radial arteries are not felt, cannulation of brachial or femoral arteries may be considered. However, these sites should be changed to more peripherally placed catheters as early as possible to avoid complications such as distal ischaemia.
- The distal circulation must be assessed periodically by capillary refill time, colour of the digit, nail bed, pulse oximetry, etc.

What information is obtained?

The invasive arterial pressure monitoring provides the following information

- **Systolic, diastolic and mean blood pressures**
- **Heart rate and rhythm**
- **Contractility:** The upstroke of the arterial waveform is steep if the heart has good contractility.
- **Preload:** The systolic variation in arterial pressure > 10% with respiration indicates hypovolaemia.
- **Afterload:** The dicrotic notch is lower on the downstroke of arterial waveform when afterload is reduced.

What size cannula?

- A 20-gauge cannula may be used for the radial, brachial or dorsalis pedis arteries in adults.
- A long 18 gauge nonkinking cannula is preferred for femoral arterial lines in adults.
- A 22-gauge cannula in children.

Complications

- Ischaemia distal to cannula: Normally collateral circulation is adequate to maintain perfusion distal to the arterial cannulation site. Occasionally ischaemia may occur when associated with low cardiac output, sepsis, shock, high-dose vasopressors or vasculitis.
- Exsanguination: In case of accidental disconnection, blood loss of up to 500 ml/min can occur (through 18 G cannula)
- Spurious result: Wrong position or calibration of transducers
- Infection
- Intra-arterial injection of drugs.

CENTRAL VENOUS PRESSURE MONITORING

A catheter inserted into the central vein with its tip positioned in the superior vena cava enables measurement of the **right atrial pressure** or central venous pressure. It also provides a good venous access.

Normal values: 2–5 cmH_2O in a spontaneously breathing patient. 5–10 cmH_2O in a mechanically ventilated patient.

Indications of central venous cannulation

1. To guide fluid therapy as in hypovolaemia, low cardiac output states, renal failure
2. For total parenteral nutrition
3. To infuse potent drugs such as antibiotics, vasopressors, chemotherapeutic agents
4. When peripheral venous access is not available
5. For PA catheterisation and cardiac output measurement.

Sites of insertion

- **Internal jugular vein** (IJV): **Right IJV is preferred** over left IJV because:
  - RIJV is more in line with the superior vena cava.
  - Injury to thoracic duct is avoided.
- **Subclavian veins** (Figs 48.22 and 48.23).
- Peripherally placed central venous catheters may also be useful in measuring CVP if the tip of the catheter is intrathoracic.
- Femoral vein catheters may also be used provided the tip of the catheter is close to the diaphragm in the inferior vena cava. However, it is not preferred as it is close to the inguinal region and maintaining asepsis is difficult. It also interferes with the patient's mobility.

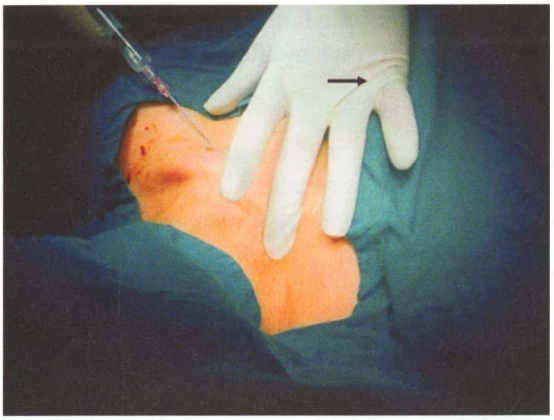


Fig. 48.22: Subclavian venous cannulation: Locating subclavian vein

Fig. 48.23: Subclavian venous cannula in place

KEY BOX 48.5

FLUID CHALLENGE TEST

CVP (cmH_2O)	Amount of fluid given over 10 min
8	200 ml
8–13	100 ml
> 13	50 ml

- If CVP rises by > 5 cmH_2O, stop fluid administration
- If CVP rises by < 5 but > 2 cmH_2O, stop fluids, wait for 10 minutes and then give a smaller volume
- If CVP rise is < 2 cmH_2O, repeat fluid administration.

CVP as a guide to fluid replacement (Key Box 48.5)

CVP may be used as a guide to right ventricular filling. The 5–2 rule is a useful way of assessing fluid responsiveness. Baseline CVP is recorded and then fluid is infused (50–200 ml of colloid over 10 min) if hypovolaemia is suspected.

Limitations

- The CVP displayed depends on correct positioning of the tip of the catheter.
- The position and calibration of the transducer are important. The transducer must be placed at the level of the midaxillary line (supine patient) and the sternal angle (sitting patient) and then zeroed to atmospheric pressure.
- It can change with changes in right ventricular compliance also. For example CVP does not reflect left ventricular filling in patients with pulmonary hypertension or in right ventricular dysfunction. In such cases, dynamic changes in CVP are more useful than absolute values.

Complications of central venous catheterisation

Associated with insertion

- Bleeding
- Carotid artery puncture
- Pneumothorax
- Haemothorax
- Air embolism
- Arrhythmias

Associated with use

- Catheter-related infection and sepsis
- Disconnection, leading to bleeding or air embolism
- Pleural or pericardial effusion due to misplaced catheter

PULMONARY ARTERIAL (PA) CATHETERISATION

These are long catheters placed in the PA through the central veins, right atrium and right ventricle (Fig. 48.24). The tip of this catheter ends in a small distal branch of the pulmonary artery. When the balloon on its distal tip is inflated, the pressure distal to the catheter reflects the **left atrial pressure** which may reflect left ventricular end-diastolic pressure and hence the preload more accurately (Fig. 48.25). PA catheter insertion may be associated with a number of complications and must be used judiciously.

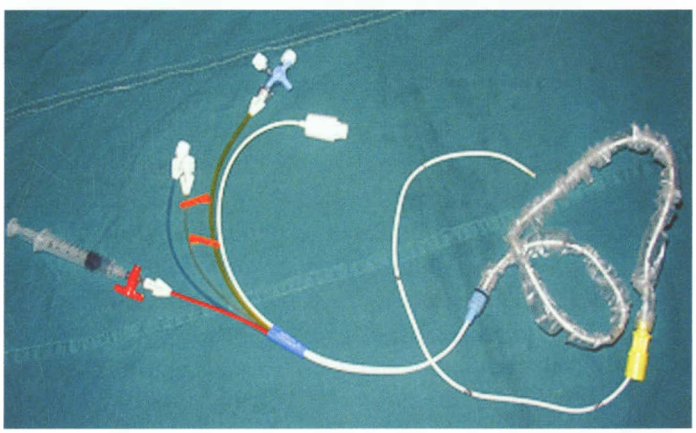


Fig. 48.24: Pulmonary artery catheter

Uses

- If a PA catheter is in place, 7–3 rule can be used to guide fluid challenge similar to the 5–2 rule with a CVP catheter.
- A rise in PCWP may be the earliest indicator of left ventricular dysfunction. A new onset mitral regurgitation can also be detected in the wedge pressure trace.
- Some PA catheters can be used to pace the left ventricle
- Cardiac output measurements are possible.

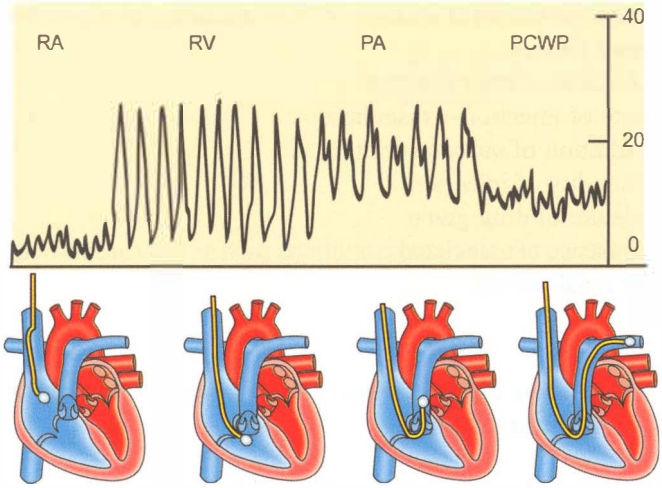


Fig. 48.25: Insertion of a PA catheter with the pressure trace display as a guide. The corresponding position of the catheter in the heart is shown diagrammatically below

LOCAL ANAESTHETICS

Local anaesthetics are drugs when injected around the nerves block impulse conduction distal to the site of injection and produce analgesia and anaesthesia in that area.

Classification

Local anaesthetics consist of a hydrophilic tertiary amine group linked to a lipophilic aromatic group. They are classified into two main categories based on this linking group: the aminoamides and the aminoesters.

- **Aminoesters:** Procaine, chloroprocaine, tetracaine
- **Aminoamides:** Lignocaine, bupivacaine, ropivacaine.

Mechanism of action

A nerve impulse is transmitted by progressive opening of sodium channels across the membrane and sudden influx of sodium into the intracellular fluid. Local anaesthetics produce **sodium channel blockade.** They block the fast sodium channels and the sodium influx, thus blocking all impulse transmission across the membrane.

Local anaesthetic exists in two forms: Ionised and nonionised. The nonionised form is lipophilic and crosses the phospholipid membrane more easily. The ionised form is hydrophilic and blocks the channel in the open state and blocks nerve transmission (use-dependent blockade). The drug blocks the channel from the intracellular direction.

Factors influencing activity

Lipid solubility

Higher the lipid solubility, higher is its ability to penetrate the lipoprotein membrane and hence greater is its potency.

pKa

The pKa of a drug is the pH at which the ionised and nonionise portions of the drug are equal. The lower the pKa, lower is th degree of ionisation for any given pH. The nonionised portio is lipophilic and crosses the cell membrane more easil hastening the onset of nerve blockade. For example, lignocain has a pKa of 7.9 and acts faster than bupivacaine with a pK of 8.1.

pH

Acidosis decreases the proportion of the nonionised drug an reduces the amount of drug able to cross the membrane. Tha is why local anaesthetics do not work optimally when injecte into an infected tissue (pH is acidic in these tissues).

Protein binding

The greater the degree of protein (membrane proteins) binding longer is the duration of action.

Choice of local anaesthetic agents

- **Lignocaine:** Skin infiltration—0.5–1%
  Minor nerve block—1%
  Epidural—1.5–2%
  Spinal anaesthesia—5% hyperbaric (heavy)
  Two topical preparations—2% lignocaine jelly and 4% lignocaine spray for the mucosal surfaces of the body.
  Note: 1% means each ml contains 10 mg of the drug, 2% to 20 mg/ml, and so on.
- **Bupivacaine:** Skin infiltration, epidural—0.25%, 0.5%
  Spinal anaesthesia—0.5%, hyperbaric (heavy).
- **Ropivacaine:** Available as 0.2% for providing postoperative analgesia, labour analgesia and as 0.75% for spinal and epidural anaesthesia and nerve blocks.

Maximum recommended doses of local anaesthetics for infiltration and blocks

- Lignocaine—5 mg/kg
- Lignocaine with adrenaline—7 mg/kg
- Bupivacaine—2.5 mg/kg
- Ropivacaine—5 mg/kg, not to exceed 200 mg for minor nerve blocks.
- Lignocaine (preservative free) when used as an anti-arrhythmic agent is given IV, in a dose of 1–2 mg/kg.

Clinical effects

Local effects

Local anaesthetics block the sodium channels in the neuronal membrane and thus the propagation of impulses across it.

Systemic effects

These drugs can produce systemic effects when high plasma levels of the drug are achieved. This may also be deliberate as when lignocaine is used as an antiarrhythmic agent. It is

classified as Class Ia drug (sodium channel blockers) in the Vaughan Williams classification of antiarrhythmic agents.

When high plasma concentrations of local anaesthetics are achieved, either due to accidental intravenous injection of the drug or due to intravascular absorption of large amount of drug infiltrated in a region, systemic toxicity can occur.

PEARLS OF WISDOM

Adrenaline containing preparations should not be used for nerve blocks of fingers, toes and penis as it can cause ischaemia.

Toxicity of local anaesthetics

Systemic toxicity

If significant amount of local anaesthetics reach the tissues of heart and brain, they exert the same membrane stabilising effect as on peripheral nerve, resulting in progressive depression of function. The toxicity of local anaesthetics is dose-dependent. These drugs always produce central nervous system (CNS) toxicity first. As the plasma level rises, cardiovascular toxicity and collapse occur.

The plasma levels of lignocaine required to produce cardiovascular collapse (CVS toxicity) is seven times that required to produce convulsions (CNS toxicity). Bupivacaine requires only three times the plasma level to produce cardiac toxicity as for CNS toxicity. Thus, bupivacaine has a greater potential for cardiotoxicity.

PEARLS OF WISDOM

With all local anaesthetics, central nervous system toxicity always comes first. This is followed by cardiac toxicity.

Clinical features of local anaesthetic toxicity are related to the plasma level of the local anaesthetic. The clinical effects and their relation to plasma level of lignocaine are given in Table 48.4.

The likelihood of toxicity of local anaesthetics depends on several factors:

1. Amount of drug injected
2. Site of injection—vascularity
3. Addition of vasoconstrictors
4. Rapidity of injection
5. Nature of drug given
6. Presence of associated conditions such as low cardiac output or renal failure.

PEARLS OF WISDOM

Bupivacaine and preservative-containing lignocaine are not for intravenous use.

Amount and nature of drug injected

Lignocaine is mostly used to produce peripheral neural conduction blockade. It can be used in doses not exceeding 5 mg/kg body weight for plexus blocks or infiltration. When combined with vasopressors such as adrenaline (1:200,000 = 5 μg/ml), the dose can be increased up to 7 mg/kg. The intravenous dose of lignocaine is 1–1.5 mg/kg when used as an antiarrhythmic. However, even a small dose such as 20 mg in an adult when injected accidentally into the carotid artery may be sufficient to produce convulsions.

Bupivacaine is used only for nerve blocks or infiltration. It is not an antiarrhythmic drug. Bupivacaine is cardiotoxic and care should be taken not to exceed the prescribed doses. Circulatory collapse and cardiac arrest due to large doses of bupivacaine can be very resistant to resuscitation. It can be used in a dose of up to 2.5 mg/kg body weight.

Ropivacaine is a newer amide local anaesthetic agent which is similar to bupivacaine but with less cardiotoxicity. A levo isomer of bupivacaine, called levobupivacaine is also less cardiotoxic and has been put into clinical use recently.

Site of injection

Certain sites are very vascular as compared to others. A higher plasma level of local anaesthetic is reached when the same amount of local anaesthetic is used for multiple intercostal nerve blocks as compared to brachial plexus block.

Table 48.4 The clinical effects and their relation to plasma level of lignocaine

Plasma	CNS toxicity	CVS toxicity
Concentration (μg/ml)		
5	Tingling, numbness, tinnitus, light-headedness.	
5–10	Slurred speech, muscle twitching	
10	Loss of consciousness	
10–15	Convulsions	
15	Coma	Myocardial depression
20	Respiratory arrest	Cardiac arrhythmias
25		Ventricular arrest

Prevention of toxicity

- Do not exceed recommended doses.
- Aspirate to rule out presence of the needle tip in a vessel before injecting the drug.
- Avoid injecting large boluses at once. Small boluses, given slowly to achieve the desired effect are safer.

Treatment of local anaesthetic toxicity

The toxicity of local anaesthetics manifests as CNS depression and convulsions. Maintenance of airway, breathing and circulation must be a priority. These convulsions generally last for a short period of time.

- Patency of the airway must be maintained.
- Oxygen by face mask.
- Ventilation, if apnoea occurs.
- Convulsions are treated with intravenous diazepam or thiopentone in incremental doses.
- Cardiovascular collapse with ephedrine, inotropes and vasoconstrictors, and CPR as needed.
- Arrhythmias must be treated appropriately.

SPINAL AND EPIDURAL ANAESTHESIA

- Injection of local anaesthetics around the spinal cord to produce a reversible blockade of impulses that pass through it, is called **central neuraxial blockade**.
- When the local anaesthetic is injected into the cerebrospinal fluid bathing the spinal cord, it is called **spinal anaesthesia** (subarachnoid block).
- When the local anaesthetics are injected into the epidural space to block the nerves that emerge from the spinal cord, it is called **epidural anaesthesia**.

SPINAL ANAESTHESIA

Physiological effects

Nervous system

The local anaesthetics spread from the site of injection by mixing with the cerebrospinal fluid (CSF). They reach the nerve fibres in the spinal cord and block transmission of impulses below the highest level of spread. The concentration of the drug is higher in the caudal regions and reduces with increasing distance cranially. A lower concentration of the drug is sufficient to block smaller fibres (C and Aδ) whereas the thick motor fibres (Aα) require a larger concentration to get blocked. Thus, **a differential blockade** is seen after a spinal anaesthetic and is as follows:

- Motor block up to a certain level (depends on the dose of drug injected)
- Sensory level of block 2 segments higher than motor block.
- Sympathetic block about two to four segments above the level of sensory block.

Cardiovascular system

- **Hypotension:** Due to the blockade of sympathetic nerve below the level of spinal block, there is profound vaso dilatation in the affected areas. A relative hypovolaemia occurs and hypotension is usually seen. The extent o hypovolaemia depends on the preoperative volume status level of block and ability to compensate for the vaso dilatation. The vessels in the upper limbs constrict to compensate for vasodilatation in the lower limbs. This is described as **pink trousers, blue jacket** phenomenon.
- **Bradycardia:** Heart rate is maintained in low blocks, but in higher blocks (high thoracic), sympathetic nerve block of the cardioaccelerator nerves can occur causing unopposed action of the parasympathetic system and bradycardia.
- **Cardiac output also reduces** in high spinal anaesthetics.

Respiratory system

- No changes in respiratory function are seen in spinal anaesthetics below T10 level.
- When the level of spinal anaesthesia ascends, the intercostal nerves are gradually blocked.
- The diaphragm is not easily paralysed as the phrenic nerve is a thick and strong nerve and arises from cervical nerve roots (C_3, C_4, C_5).
- In high spinals, the alveolar ventilation reduces and may lead to hypoxia and hypercarbia.

Gastrointestinal system

- Unopposed parasympathetic activity leads to constriction of gut with increased peristaltic activity.
- Nausea, retching or vomiting may occur and may be the symptom of impending hypotension. These symptoms disappear when the hypotension is corrected. Occasionally, it may need administration of an anticholinergic or an antiemetic agent.
- However, since the bowel is contracted and small and the skeletal muscle relaxation produced is greater, the surgeons find it easier to operate on such a gut.

Indications

- Any surgery below the level of umbilicus

Contraindications

Absolute

- Patient refusal
- Infection at the site of injection
- Bleeding tendencies

Relative

- Hypovolaemia
- Severe stenotic valvular heart disease

Limitation
Limited duration of block

Preprocedure check
A preprocedure check of the anaesthesia equipment, resuscitation equipment and drugs is made. An intravenous line is placed and monitoring commenced. The patient is then positioned for spinal anaesthesia.

Position (Figs 48.26 and 48.27)
The spinal anaesthetic may be administered with the patient in lateral or sitting position.

Lateral: The patient lies either in the left or right lateral position. The back should be parallel to the edge of the operating table and perpendicular to the ground. The legs should be flexed at the hips as much as possible.

Sitting position: The patient sits on the table, with the back bent forward. He is allowed to rest his arms on pillows. The back is cleaned with spirit and betadine and draped. Under aseptic precautions, the vertebral spines are identified in the lumbar region. The highest point of the iliac crest corresponds

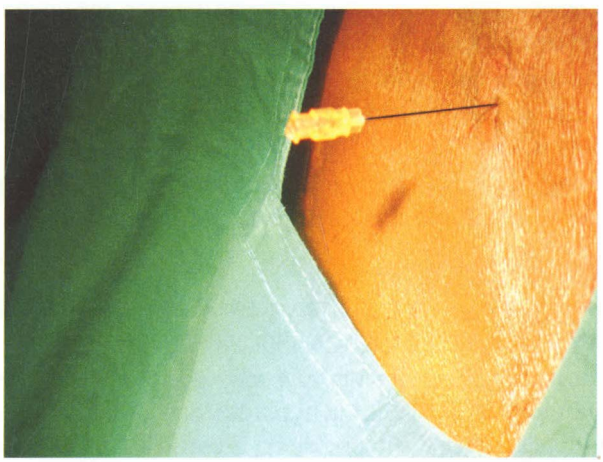


Fig. 48.26: Spinal anaesthesia in the lateral position, spinal needle is in place and CSF drop is seen at the hub

to L3–4 space. The L2–3, L3–4, L4–5 intervertebral spaces can also be used. A space higher than this is not used as the spinal cord ends at L1 in adults. This point is lower in children and should be borne in mind in paediatric spinals.

Approach
The subarachnoid space may be approached either from the midline or by a paramedian technique. A subcutaneous wheal of local anaesthetic is raised in the chosen intervertebral space.

Midline approach: The lumbar puncture needle is inserted in the midline, midway between the spines and perpendicular to the skin. The spinal needle passes through the following structures to reach the subarachnoid space:

1. Skin
2. Subcutaneous tissue

Fig. 48.27: Spinal anaesthesia in the sitting position

3. Supraspinous ligaments
4. Interspinous ligaments
5. Ligamentum flavum
6. Dura and arachnoid

Paramedian approach: The needle is inserted a finger-breadth lateral to the spine and advanced in a slightly cephalad direction towards the midline. If the needle touches the lamina, it should be redirected medially. The correct position of the needle is identified by obtaining a free flow of CSF. This approach helps access the subarachnoid space in those patients whose interspinous and supraspinous ligaments are **calcified or in patients unable to bend** enough to open the interspaces well.

The local anaesthetic is now injected into the CSF, taking care not to displace the needle.

Complications
These may be classified into **Minor** and **Major based on the reversibility and seriousness of the complication.**

Minor

Hypotension
This is treated with intravenous fluids to compensate for the vasodilatation. If necessary, incremental doses of a vasoconstrictor may also be used.

Bradycardia
If the cardioaccelerator nerves (T1–T4) are blocked. This is usually easily treated with an anticholinergic such as atropine or glycopyrrolate. If profound, a small dose of adrenaline may be required (very rare).

Postdural puncture headache (PDPH)
The incidence of PDPH depends on the size of the needle used, number of punctures made, fluid status and ambulation. With finer (25 and 26# needles) and good hydration of the patient,

PDPH is uncommon. This may be treated with rest, increased fluid intake, plenty of coffee and NSAIDs. Rarely, an epidural blood patch *(vide infra)* may be required.

Respiratory depression

If the level of spinal anaesthesia is high and all intercostal muscles are paralysed, respiratory depression may occur. However, diaphragm, the principal muscle of respiration is supplied by the thick phrenic nerve which does not get blocked easily. Any respiratory depression seen during spinal anaesthesia is more due to hypoperfusion of the respiratory centre (due to hypotension). This can be treated with respiratory support as required and stabilisation of blood pressure.

Retention of urine

Backache: This is not a problem of spinal anaesthesia *per se* but may be due to faulty positioning during surgery.

Major

1. **Infection:** Arachnoiditis, meningitis
2. **Nerve injury:** Cauda equina syndrome

EPIDURAL ANAESTHESIA (Table 48.5)

In this type of central neuraxial blockade, local anaesthetic is injected in the space around the dura (epidural space). The local anaesthetic blocks the nerves as they emerge through the intervertebral foramen. Some of it diffuses through the meninges into the spinal cord and acts on the spinal cord.

Preprocedure check

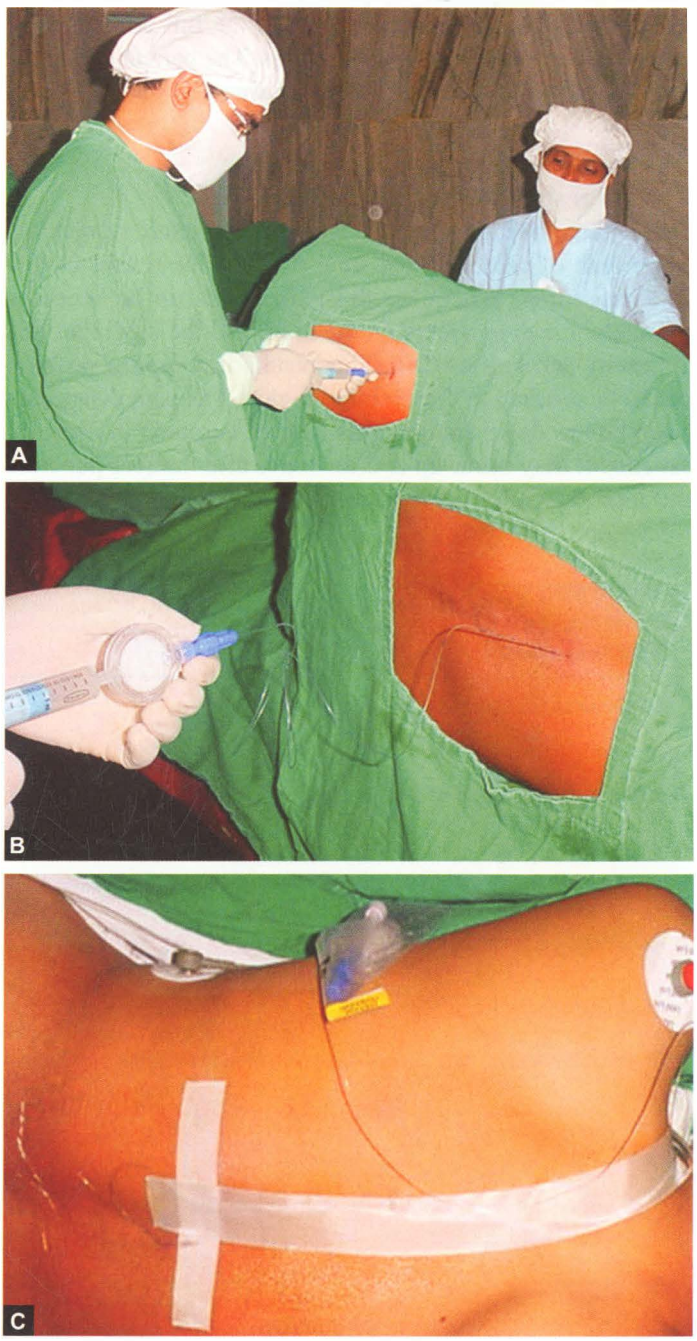
A preprocedure check of the anaesthesia equipment, resuscitation equipment and drugs is made. An intravenous line is placed and monitoring of heart rate, electrocardiogram, blood pressure and oxygen saturation is established and the baseline noted.

Position

The epidural puncture can be done with the patient in sitting position or in the lateral decubitus position.

Technique

The patient's back is cleaned with an antiseptic and then draped. Under aseptic precautions, epidural puncture is done using a 16 or 18# Tuohy needle. This needle has a **blunt tip** to reduce the risk of dural puncture. The needle is inserted either in the **midline** or by a **paramedian** approach. It passes through the same tissues as in lumbar puncture except the subarachnoid space. The needle is inserted along with its stylet and always advanced slowly from skin onwards. Once the subcutaneous tissue is entered, the stylet is removed. A 2 ml or 5 ml syringe with a freely moving plunger and containing either air, saline or both is then connected to the needle hub. A gentle attempt at injection of this air or saline is made as the needle advance through the tissues. The entry of the needle into the epidura space is heralded when it penetrates the ligamentum flavur and there is a **loss of resistance to injection of air or saline** This is taken as the end-point. An epidural catheter is passe through this needle and advanced to about 3–4 cm into th space. The needle is removed and the catheter taped and fixe to the back. A bacterial filter is attached to the injection por of the catheter (Figs 48.28A to C).



Figs 48.28A to C: Epidural anaesthesia: (A) Eliciting loss of resistance to air, (B) Epidural catheter attached to bacterial filter, (C) Epidural catheter taped to the back

The epidural needle or catheter may accidentally enter an epidural vein or the subarachnoid space. To avoid injecting a large dose of local anaesthetic into either of these spaces, a test-dose containing a small amount of local anaesthetic (3 ml of 2% = 60 mg lignocaine) and 15 µg of adrenaline is injected. Any sensory or motor block following this dose would suggest an accidental dural puncture resulting in spinal anaesthesia. An accidental intravascular injection is identified by an increase in the heart rate and blood pressure within a minute of the injection. In either situation, the epidural catheter may need to be withdrawn or replaced. If neither response is seen, an epidural placement is assumed and the full dose of local anaesthetic is injected in divided doses. The patient should be continuously monitored till the block wears off.

Complications

1. Postdural puncture headache (PDPH)

The epidural needles are large and dural puncture results in a larger leak of cerebrospinal fluid (CSF). This results in low CSF pressures. Whenever the patient sits up or becomes ambulatory, a drag occurs on the brain and the meninges due to gravity and loss of CSF. This results in a typical postural headache referred to the occipital region. The pain disappears when he lies down supine. This is more common in obstetric patients. It may occur up to 2 to 7 days after lumbar puncture and may persist for up to 6 weeks.

Treatment

Plenty of oral fluids may increase CSF production. Rest, plenty of coffee and NSAIDs may also help. Rarely, an epidural blood patch may be required.

Epidural blood patch: If the headache is very severe, an epidural blood patch may be given. 10 to 15 ml of patient's own blood is drawn under aseptic precautions. Simultaneously, epidural puncture is made in the same space as the previous epidural puncture. The freshly drawn blood is injected into the epidural space which clots and seals the puncture hole. This is nearly 100% effective in relieving the headache.

2. Total spinal block

When a large dose of local anaesthetic is injected intrathecally inadvertently, all spinal nerves are blocked, causing profound hypotension, bradycardia and collapse. If the patient is continuously monitored and treated promptly, this is completely reversible.

Treatment

1. Volume infusion and vasopressors
2. Endotracheal intubation and ventilation as necessary.

3. Urinary retention

4. Meningitis, if aseptic precautions are not followed.

5. Cauda equina syndrome, adhesive arachnoiditis: Extremely rare.

Table 48.5 Comparison of spinal and epidural anaesthesia

Spinal anaesthesia	Epidural anaesthesia
1. Done in the lumbar region only	Can be done in the lumbar, sacral (caudal), thoracic or cervical regions.
2. Confirmation of correct placement of needle by ensuring free flow of CSF	Placement confirmed by using loss of resistance to injection of air, saline or both
3. A small amount of local anaesthetic is used	Larger mass of local anaesthetic is injected
4. 23–29 # LP needles used	Larger needles (16–20 #) are required
5. Test doses not required	Use of test doses advisable
6. Onset of neural blockade is fast. So also side-effects	Onset slow as drugs have to penetrate the dura. So, less hypotension
7. All the nerves are blocked below the level of anaesthesia	The local anaesthetic spreads both caudad and cephalad. Segmental block can be achieved
8. Limited duration. Continuous spinals are not routinely used.	Epidural catheters are routinely introduced and hence duration of anaesthesia can be prolonged with repeated boluses or continuous infusion through catheters
9. Postdural puncture headache possible	PDPH not seen, unless dura is inadvertently punctured
10. No such problem	Inadvertent intrathecal or intravascular injection of large amount of drugs possible, with consequent complications such as total spinal blockade and local anaesthetic toxicity respectively.

OTHER REGIONAL TECHNIQUES

CAUDAL ANALGESIA

This is a very popular technique in providing postoperative analgesia in children. It involves injection of local anaesthetics with or without opioids in the caudal epidural space.

Procedure

Position

The patient is positioned in lateral position with the knees flexed and the back perpendicular to the ground. It can also be given with the patient in prone position. The area over the sacrum and the gluteal region is cleaned and draped.

Needles

A 22 or 23 G hypodermic needle or a scalp vein set is used to administer the block.

Technique

The needle is inserted at the apex of the sacral hiatus at a 60° angle to the skin. A distinct 'pop' or a 'give way' is felt as the needle punctures the sacrococcygeal membrane. The angle of the needle is then changed to about 15° to 20° to the skin and advanced a little further into the sacral epidural space. The latter step is optional and has to be done with caution as the dural sac may end relatively low in infants. After careful aspiration to rule out blood or CSF, a small dose of local anaesthetic is injected. There should be no resistance to injection. A subcutaneous injection should also be ruled out.

Drugs

0.25% bupivacaine in a dose of 0.5 ml/kg is sufficient for perineal and low sacral procedures, 1 ml/kg for lumbosacral procedures and 1.5 ml/kg for lower abdominal procedures. However, a total volume of 20 ml and a total dose of 2.5 mg/kg of bupivacaine may not be exceeded.

Indications

- Postoperative pain relief in children for perineal and lumbosacral procedures.
- It is also used to supplement general anaesthesia for perianal procedures in adults.

Contraindications

- Absence of consent from parents/patients
- Local infection
- Bleeding tendencies.

Complications

- Intrathecal injection is possible, especially in smaller infants who have extension of dural sac down to S3.
- Intravascular injection of large dose of local anaesthetics.

BRACHIAL PLEXUS BLOCK

Injection of local anaesthetics injected around the brachial plexus produces analgesia and even surgical anaesthesia in the upper limb. The brachial plexus can be blocked by four different approaches: interscalene, supraclavicular, infraclavicular or the axillary. Of these, the supraclavicular and the axillary techniques are the most popular.

Supraclavicular brachial plexus block

Position

The patient is positioned in supine position with the head turned to the opposite side of the block. The area over the lower part of the neck is cleaned and draped.

Needles

A 22 G hypodermic needle, a scalp vein set or an insulated needle with an internal nerve stimulator may be used to administer the block. Ultrasound guidance is becoming increasingly popular to guide nerve blocks.

Technique

The needle is inserted at a point 1 to 1.5 cm above the midpoint of the clavicle. The subclavian arterial pulsations are felt with the thumb of one hand and the needle advanced posterior to it with a medial and caudal direction. Paraesthesia must be sought in the upper limb and the injection of local anaesthetic made at the point of paraesthesia. Undue pain during injection suggests intraneural injection. Careful aspiration for absence of blood to rule out intravascular injection is mandatory before injection of the local anaesthetic.

Drugs

- Lignocaine plain not exceeding 5 mg/kg
- Lignocaine with adrenaline 7 mg/kg or
- Bupivacaine not exceeding 2.5 mg/kg may be used.

Indications

- Intraoperative analgesia and postoperative pain relief in adults and children.
- Sole anaesthetic in adults for procedures on the upper limb.

Contraindications

- Absence of consent
- Local infection
- Bleeding tendencies.

Complications

- Haematoma
- Intravascular injection of local anaesthetics
- Pneumothorax.

AXILLARY BRACHIAL PLEXUS BLOCK

Position

The patient is positioned in supine position with the arm abducted to a right angle and the elbow bent at right angles to the arm. The area over the axilla is cleaned and draped.

Needles

A 22 G hypodermic needle, a scalp vein set or an insulated needle with an internal nerve stimulator may be used.

Technique

- The needle is inserted at a point just above the point of maximum pulsations of the axillary artery but parallel to the artery. This point should be as high in the axilla as possible. Paraesthesia must be sought in the upper limb and the injection of local anaesthetic made at the point of paraesthesia. Undue pain during injection suggests intraneural injection. Careful aspiration for absence of blood to rule out intravascular injection is mandatory before injection of the local anaesthetic.
- A single injection of the local anaesthetic can be made at this point. Alternately, half the total volume of injection can be made at a point on the other side of the artery.

Drugs

Either bupivacaine not exceeding a total dose of 2.5 mg/kg; lignocaine plain 5 mg/kg or with adrenaline 7 mg/kg may be used.

A volume 35–40 ml may be required.

Indications

Intraoperative analgesia, anaesthesia and postoperative pain relief in children and adults for procedures on the upper limb.

Contraindications

Absence of consent, local infection, bleeding tendencies.

Complications

Haematoma, intravascular injection of local anaesthetics

ANKLE BLOCK

This is a popular technique in providing intra- and post-operative analgesia in adults undergoing procedures on the foot.

Position

The patient is positioned supine. The foot is raised by an assistant and the area around the ankle is cleaned and draped.

Needles

A 22 G hypodermic needle is used to administer the block.

Technique

Ankle block involves blocking of **5 nerves**.

1. The **posterior tibial nerve** is blocked with 3–5 ml of local anaesthetic at a point midway between the medial malleolus and the heel, just behind the posterior tibial arterial pulsations.
2. The **sural nerve** may be blocked at a point midway between the lateral malleolus and the heel, just lateral to the Achilles' tendon.
3. The **deep peroneal nerve** is blocked at a point midway between the lateral and the medial malleoli lateral to the tendon of extensor hallucis longus and anterior tibial artery.
4. The **saphenous nerve** and the **superficial peroneal nerves** are easily blocked by raising a subcutaneous wheal of local anaesthetic between the malleoli anteriorly.

Drugs

Lignocaine plain not exceeding 5 mg/kg or bupivacaine not exceeding 2.5 mg/kg may be used.

Indications

Postoperative pain relief in adults for procedures on the foot.

Contraindications

Absence of consent from patients, local infection, bleeding tendencies.

PAIN AND ITS RELIEF

Introduction

One of the main anxieties of undergoing an operation is the pain that is anticipated along with it. An anaesthesiologist's responsibilities do not end with providing safe perioperative care. He must also ensure that the pain associated with the surgery is relieved adequately so that the surgery itself is not so unpleasant an experience.

Definition

Pain is defined as an unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage. Pain can be **acute** or **chronic**. The pain that is seen immediately after surgery is acute postoperative pain and is easier to treat. The mechanism of perpetuation of chronic pain is less well understood and is more difficult to treat.

Pain pathways

Pain begins with injury to the tissue. Both the peripheral and the central nervous systems are involved in the ultimate pain perception (Fig. 48.29).

ACUTE PAIN

Four basic mechanisms are involved in the appreciation of pain: transduction, transmission, modulation and perception. Pain relief can be achieved by blocking each of these steps.

Transduction

The transduction of noxious stimuli begins with peripheral nociceptors. These nociceptors convert the stimulus (transducer) of injury (heat, pressure, mechanoception) to pain sensation. This is also assisted by the production of prostaglandins and leukotrienes in response to the injury. Nonsteroidal anti-inflammatory analgesics can be used to reduce their production. Local application of local anaesthetics also reduces transduction.

Transmission

Signals from these nociceptors travel along myelinated Aδ and unmyelinated C fibres. Their axons synapse in the dorsal horn of the spinal cord, where the neurons of laminae (of Rexed) I, II and V are most involved in the perception of pain. These fibres can be blocked at the nerve plexus level using local anaesthetics.

Modulation

Large fibre inputs from other sensory modalities and descending pathways can modulate activity in the dorsal horn. The extent of this modulation results in differing levels of pain experienced by patients. This can be further modified using local anaesthetics (epidural anaesthesia), opioids and alpha-2 agonists.

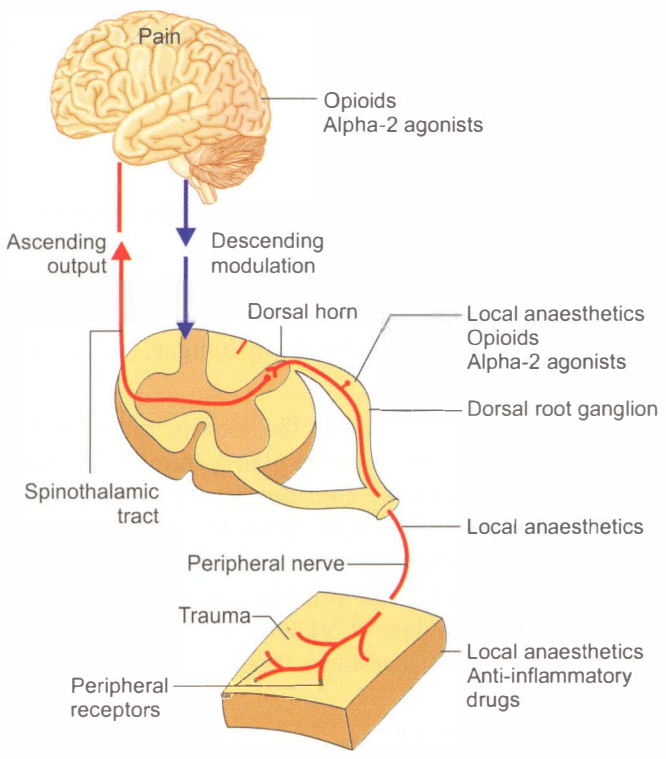


Fig. 48.29: Pain pathways and their modulation using drugs

Perception

The signals then travel along the spinothalamic tract of the spinal cord to the thalamus and the cortex which ultimately cause activity in the appropriate portion of the sensory cortex and the limbic system. The response of the cerebral cortical cells and the limbic cells can be modified using opioids and alpha-2 agonists.

Often, a combination of all these techniques is used to achieve good pain relief. Relief of pain helps the patient heal faster and return to normal activities early. The complications of inadequate pain relief such as respiratory infections, hypertension, myocardial infarction and increased catabolism are avoided or minimised.

Assessment of pain

Pain is more difficult to measure as compared to other parameters such as temperature or heart rate. The measurement of pain is mainly dependent on the patient's perception of it and may be done in any of the following ways:

1. **Visual Analogue Scale (VAS):** The pain is assessed along a 100 mm scale where 0 corresponds to no pain at all and 100 to the worst imaginable pain. The patient scores his pain at any point he finds appropriate. The pain relief is assessed thereafter from this point. The VAS is the most popular tool used to assess pain and its relief.

Fig. 48.30: Wong–Baker faces pain rating scale for children

2. **Category rating scale:** The different ratings are no pain, mild, moderate or severe.
3. **Wong–Baker Faces pain rating scale:** A scale with different faces, a happy face at one end and a face in severe pain and crying at the other with four other faces in between may be used for children (Fig. 48.30).
4. **Biological measures** such as tachycardia, hypertension, tachypnoea or inadequate breathing are relatively inaccurate but useful estimates of pain.

CHRONIC PAIN

The initial response to a noxious stimulus is brief and correlates with the sharp, well-localised initial pain. The second phase of the response is more prolonged and correlates with the dull, diffuse pain experienced after the initial injury. This second phase is associated with a growing region of hypersensitivity

around the point where the noxious stimulus was initially applied and is said to result in chronic pain. The response to noxious stimuli can be modulated by their repeated application. Peripheral nociceptors become more responsive with the repeated application of noxious stimuli. Their sensitivity can be further enhanced by many tissue factors and inflammatory mediators released in the course of tissue injury.

The process through which the neurons of the dorsal horn of the spinal cord become sensitised by prior noxious stimuli is often referred to as "windup" or "central sensitisation." Much less is known about pain-induced sensitisation of the supraspinal components of the CNS. This phenomenon leads to chronic pain where the patient experiences pain, often, excruciating, and long after the stimulus that initially caused the pain is removed. The relief of chronic pain is more difficult. Various techniques used to treat chronic pain are as follows:

Nonpharmacological techniques

- Transcutaneous electrical nerve stimulation (TENS)
- Acupuncture

Pharmacological techniques

- Paracetamol
- Nonsteroidal analgesics

Sympathetic blockade

- Sympathetic ganglion blocks—stellate ganglion block, coeliac plexus block, etc.
- Epidural local anaesthetics
- Epidural neurolytic such as phenol, alcohol injections.

COMPLICATIONS IN ANAESTHESIA

The practice of anaesthesia has become very safe due to better preoperative evaluation and preparation, careful choice of patients, better monitoring, availability of safer drugs and safer anaesthetic techniques. The incidence of complications has come down drastically. However, complications can still occur. The perioperative (pre-, intra-, and postoperative periods) complications can be classified as follows:

Respiratory

- Airway obstruction
- Bronchospasm
- Respiratory failure.

Cardiovascular

- Hypertension
- Hypotension
- Arrhythmias
- Shock
- Cardiac arrest.

Central nervous system

- Postoperative drowsiness
- Postoperative nausea and vomiting
- Neurologic complications of regional blockade.

Renal and hepatic failure

RESPIRATORY COMPLICATIONS

Airway obstruction

Airway obstruction may occur during the induction of general anaesthesia. When a person becomes unconscious, the tongue and the epiglottis fall back and can obstruct the airway. The patency of the airway is usually maintained fairly easily by the anaesthesiologist by using chin lift or jaw thrust manoeuvres. A definitive airway such as an endotracheal tube may then be inserted into the trachea.

Occasionally, the chin lift or jaw thrust manoeuvres are inadequate to maintain a patent airway as in patients with abnormal airways. Insertion of an endotracheal tube may also prove to be difficult in certain individuals. An oral or nasopharyngeal airway may be inserted to overcome this obstruction and maintenance of the airway for a short duration. If there is difficulty in inserting the endotracheal tube due to supraglottic causes and oral or nasopharyngeal airways are not sufficient to relieve the obstruction, insertion of a laryngeal mask airway or a Combitube® may be attempted. If the problem is at the glottis or subglottis and the airway is obstructed, an emergency cricothyrotomy or a tracheostomy may be required.

Perioperative airway obstruction may also be due to any of the following causes

- Trauma—maxillofacial, head injury
- Foreign body aspiration
- Laryngospasm
- Infection—Ludwig's angina, retropharyngeal abscess
- Oedema—laryngeal/pharyngeal oedema
- Neurological—recurrent laryngeal nerve injury
- Endocrine—thyroid enlargement
- Tumour—malignancy of the airway (tongue, cheek, larynx or the pharynx)

A thorough assessment of the airway must be done preoperatively and a management plan A formulated. Plan B and Plan C also should be considered in the eventuality of failure of Plan A. Generally loss of life occurs not because of inability to intubate but due to an inability to oxygenate and ventilate the patient. This situation, also called, 'cannot intubate—cannot ventilate' (CVCI) is one of the most dreaded situations faced by an anaesthesiologist.

Bronchospasm

Bronchospasm may occur in a patient under anaesthesia. The possible causes are as follows:

- Irritable airways as in a known asthmatic, chronic obstructive airways disease, i.e. exacerbation of pre-existing bronchospastic disease.
- Endobronchial intubation and cranial stimulation
- As part of allergic reaction to anaesthetic drugs, antibiotics
- Aspiration of regurgitated gastric contents
- Pneumothorax
- Upper airway obstruction and laryngospasm can reflexly stimulate bronchospasm.

Treatment involves treatment of the precipitating cause and bronchodilators.

Respiratory failure

A patient is said to be in respiratory failure if he is unable to maintain adequate oxygenation and ventilation, i.e. arterial blood gas tension of O_2 less than 60 mmHg (when patient is breathing 60% O_2) and CO_2 more than 50 mmHg. This could be acute or acute exacerbation of chronic respiratory failure.

Causes

Central

- Head injury
- Depressant drugs—anaesthetics, opioids
- Hypoxic encephalopathy
- Metabolic causes such as hyponatraemia, hypoglycaemia, hypokalaemia, hyperglycaemia.

Peripheral

- Lung parenchymal disorders such as pneumonia, atelectasis, aspiration, pneumothorax, pulmonary embolism
- Inadequate respiratory excursion due to pain or thoracic cage abnormalities such as kyphoscoliosis.
- Weakness of muscles, e.g. prolonged effect of muscle relaxants, myasthenia gravis

Treatment

- Treat the cause
- Intermittent positive pressure ventilation and ventilatory support till the patient improves.

CARDIOVASCULAR COMPLICATIONS

Hypertension, hypotension and arrhythmias occur perioperatively due to various reasons such as inadequate preoperative treatment, surgical stress, inadequate anaesthesia, metabolic or endocrine causes or even drug interactions. Brief periods of haemodynamic instability, although common are well-tolerated by healthy individuals. Continuous and appropriate monitoring and prompt treatment should avoid long-term complications.

Hypotension may be due to decreased preload, reduced contractility or increased afterload to the left ventricle. If not identified or treated in time, it may progress to shock and cardiac arrest. Hypovolaemic shock is the commonest type of shock encountered during surgery. However, cardiogenic shoc due to perioperative myocardial infarction, anaphylactic shoc due to allergic reaction to anaesthetics or neurogenic shoc due to vasovagal attack, high spinals may also occur i susceptible individuals.

Perioperative myocardial infarction (MI) is a compl cation that occurs in susceptible individuals. It may be cause due to extreme and sustained variations in haemodynamic such as hypotension, hypertension or tachycardia. It may als be caused due to thrombosis as the patient is in hypercoagulable state due to stress of surgery. The highe incidence of perioperative MI is seen not on the day of surger (when the patient is under the vigilant care of th anaesthesiologist) but on the second or the third day when th attention given to him is less in terms of pain relief o haemodynamic changes.

CENTRAL NERVOUS SYSTEM COMPLICATIONS

Awareness

Rarely, a patient under GA might recall events that occurre during the procedure. This is termed awareness durin anaesthesia and is one of the most dreaded complications b both anaesthesiologist and the patient. This is more likely t occur if the patient's haemodynamics are very unstable (a in postpartum haemorrhage, trauma) and the anaesthesiologis fears further cardiac depression may occur with the use o inhalation agents. The use of opioids and nitrous oxide ma provide analgesia but not the amnesia and anaesthesia required With increased awareness of this complication amon anaesthesiologists coupled with the use of benzodiazepine and modern anaesthetic agents which are more cardiostable the incidence of this is reduced. A new monitor called BI index monitor provides some information of the conscious state of the patient but is not widely available yet.

Postoperative drowsiness

A patient may be slow to awaken after anaesthesia due to persistent effect of the anaesthetic agents or opioids administered during the anaesthesia. However, it may be due to metabolic causes such as hypoxia, hypothermia, hypo- or hypernatraemia, hypo- or hyperglycaemia. If all these causes are ruled out, a neurological consultation is obtained to rule out any space-occupying lesions in the central nervous system, stroke or hypoxic encephalopathy.

Postoperative nausea and vomiting (PONV)

PONV is a frequent complication of anaesthesia. It is common in women, after laparoscopic surgeries, squint surgeries and is associated with the use of nitrous oxide and opioids or even early oral intake postoperatively. It may be treated with IV metoclopramide (10 mg), ondansetron (4–8 mg) or dexamethasone (4–8 mg) in an average adult.

Nerve injuries

Nerve injuries may arise as a complication of regional blockade. Complications such as adhesive arachnoiditis, cauda equina syndrome or paraplegia have been reported after spinal and epidural anaesthesia but are extremely rare. The use of tourniquet, if prolonged or if very high pressures are used, can also cause nerve injuries.

Peripheral nerve injuries may occur perioperatively due to improper positioning under regional or general anaesthesia. The common peroneal nerve and the sciatic nerve can be injured during lithotomy position. The ulnar and the radial nerves may be affected in the arm or at the elbow due to inadequate attention to positioning. Brachial plexus stretch injury can occur if the arms are allowed to be abducted more than ninety degrees. Injury to optic nerves or the retina may occur in prone position due to compression of the eyeball. Corneal injury may occur due to exposure in an unconscious patient.

RENAL FAILURE

Renal failure, usually prerenal is associated with large fluid shifts or major haemodynamic instability. Direct injury to the kidneys (acute tubular necrosis) may occur if adequate attention is not given to prerenal failure. Methoxyflurane, an inhalation anaesthetic agent can cause renal failure but is no longer in clinical use. Renal failure can also occur as part of hepatorenal syndrome or after a mismatched blood transfusion.

HEPATIC FAILURE

A patient with compromised hepatic function may proceed to hepatic failure perioperatively, e.g. cirrhosis of liver, obstructive jaundice. Hepatic failure may also occur due to infective complications such as hepatitis or sepsis. Massive hepatic necrosis has been reported after repeated use of halothane. The incidence of this is very rare and must be a diagnosis of exclusion.

CARDIOPULMONARY RESUSCITATION (CPR)

Cardiac arrest is an inevitable and natural event in every one's life. However, this event may have been precipitated prematurely due to some underlying but treatable cause. If prompt intervention is made and the patient's heart and respiration are supported at this stage, precious time may be gained to treat this precipitating cause. Many victims of cardiac arrest may lead normal lives once this calamity is tided over. This is the basis of cardiopulmonary resuscitation.

CPR when given without the help of any equipment is called Basic CPR (also called basic life support—BLS). When equipment and drugs are used in CPR, it is called Advanced CPR (advanced cardiac life support—ACLS). CPR must be given promptly and in the correct manner to be effective. While basic CPR alone may not be sufficient in resuscitating a victim of cardiac arrest, it is this intial response that keeps the patient alive till advance help can be provided. Success of resuscitation as well as health-related quality of life depends largely on the promptness and quality of basic CPR. Hence, knowledge of CPR is mandatory for all medical and paramedical personnel.

Points to note

- CPR is only a symptomatic therapy. Attention must be given to the precipitating cause and must also be treated.
- The best of CPR can deliver only one-third of the normal cardiac output. Hence, it is important to aim for return of spontaneous circulation as early as possible.
- 'Time is brain': Effectiveness of advanced cardiac life support as well as intact survival largely depends on prompt, early and good quality basic CPR.

Respiratory arrest

The patient stops breathing due to a primary respiratory cause. The heart continues to beat till all the oxygen in the lungs is removed. The heart will stop once hypoxia occurs. This may take 1–2 minutes to occur. If the respiration is assisted promptly, it is possible to avert cardiac arrest. However, this is possible only in a witnessed respiratory arrest.

Cardiac arrest

The heart stops beating and there is no circulation. Oxygen delivery to the tissues stops. Within 15 seconds of a cardiac arrest, a person loses consciousness. The brain stops functioning within 3 minutes. This is called **survival time.** If resuscitation is not done in another five minutes, brain death occurs. This is called **revival time.** Many factors influence these survival and revival times. The important 'take home' message is that **prompt and effective treatment is necessary for successful resuscitation.**

BASIC CPR

The sequence of steps of basic CPR has been changed in the 2010 guidelines of American Heart Association (AHA). Approach to CPR is currently done in the following order: C, A and B. C for cardiac compressions, A for airway and B for breathing.

There are six steps in basic CPR.

STEP 1: ASSESS UNCONSCIOUSNESS. "SHAKE GENTLY AND SHOUT LOUDLY"

In cardiac arrest, cerebral circulation comes to a standstill and the person becomes unconscious within 15–20 seconds. Thus, a person in cardiac arrest will not be conscious. Shortly thereafter, the victim of cardiac arrest will also stop breathing. On shaking gently and calling out loudly, if there is no response, assume unconsciousness. Turn the patient in to a supine

position. Take care to turn the head, neck and body together in a trauma victim who may possibly have a cervical spine injury. Observe for any sign of breathing such as chest movement. If there is no breathing, it confirms that the patient needs assistance.

STEP 2: CALL FOR HELP AND AED

The chances of resuscitation are lower with basic CPR and it is important to get help (in the form of ambulance services for advanced CPR) as early as possible. It is also difficult for a single rescuer to administer CPR for long periods. The rescuer may take up to a minute to call for help. Since the majority of adult, nontraumatic cardiac arrests are due to ventricular fibrillation or pulseless ventricular tachycardia (shockable rhythms), a defibrillator, either manual or automated, must be obtained as quickly as possible.

STEP 3: FEEL CAROTID PULSE

Check for pulse or other signs of circulation such as coughing, movement in response to the rescue breaths. When circulation is inadequate, the body tries to conserve circulation to the vital organs as long as possible. Thus, the carotid artery pulsations are the last to disappear when cardiac output gradually reduces. Hence, it is important to feel the carotid pulse to identify cardiac arrest (Fig 48.31). Infants and neonates have short necks and it is difficult to feel their carotid arteries. In them, the brachial artery may be used to identify cardiac arrest. If no pulse is felt, the patient is in cardiac arrest.

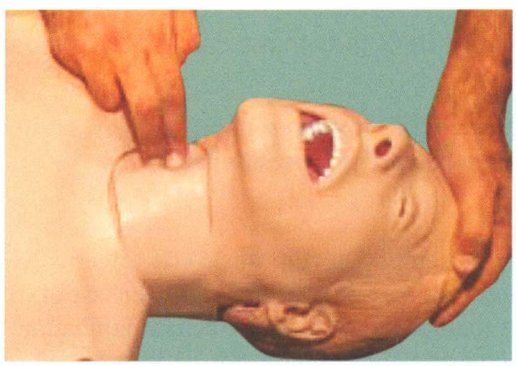


Fig. 48.31: Feel carotid pulse

STEP 4: GIVE CHEST COMPRESSIONS

Site of compression

Compressions should be given with the heel of the hand placed on the lower half of the sternum. Care should be taken not to compress on either side of the sternum as it may cause rib fractures.

Technique (Fig. 48.32)

Place the heel of one hand over the lower half of the sternum in the midline. Clasp this hand with the heel of the other hand, keep the elbows straight and move forward so as to place the shoulders directly above the sternum. The rescuer can avoi fatigue during compressions by allowing his or her body weigl to be transmitted to the chest to create the required depth c compression rather than use muscle power. Thus, th movement occurs at the rescuer's hips and not at the elbow Only one hand may be used for compression in children les than 8 years of age.

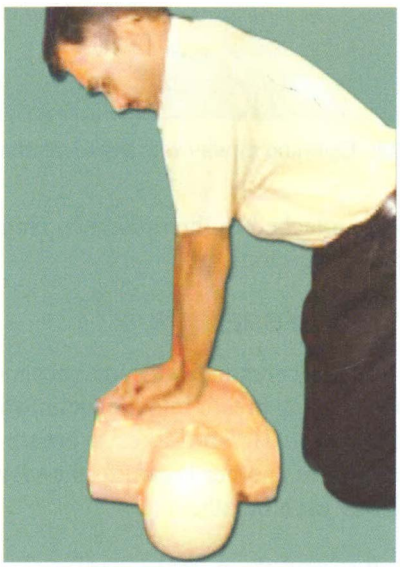


Fig. 48.32: Provide chest compressions

Rate of compression: Provide 30 compressions at a rate of a least 100/minute.

Depth of compression: 2 inches (5 cm) in an adult.

Chest compressions are the most important part of basic CPR. Hence they should be initiated as soon as cardiac arrest is recognised. 'Push hard, push fast' is what the AHA recommends. There should be minimal interruption in chest compressions for any other interventions such as defibrillation, airway management or even calling for help. The patient should be lying on a relatively hard surface. If not a hard board may be placed underneath the patients chest so that the compressions become effective.

STEP 5: OPEN AIRWAY

When a person loses consciousness, the muscles of the tongue, epiglottis and pharynx relax. The tongue and the epiglottis fall back and obstruct the glottis. Unless the airway is opened, it is not possible to check accurately whether respiration is present or not and if present, whether it is adequate or inadequate. The airway is opened using a "head-tilt and chin lift method" (Fig 48.33).

The palm of one hand is placed on the forehead of the patient and the head tilted posteriorly to extend the head as much as possible. The chin of the patient is lifted up manually with the thumb and fingers of the other hand so that the lower incisors of the victim override the upper incisors. Since the tongue and

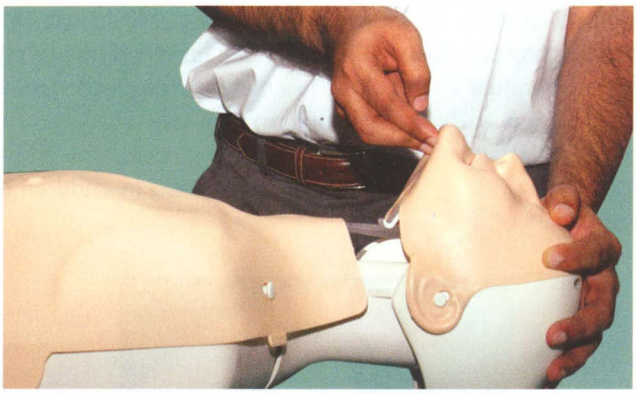


Fig. 48.33: Opening airway with 'head tilt and chin lift'

the epiglottis are attached to the mandible, chin lift also lifts the tongue and the epiglottis.

STEP 6: GIVE TWO BREATHS

The rescuer opens his/her mouth wide, makes a mouth to mouth seal and delivers a large breath into the victim so as to expand the chest adequately. He/she takes a breath in from the atmosphere and gives another breath to the victim. Take two seconds to deliver each breath.

Compression-ventilation ratio

Provide cycles of 30 compressions and 2 breaths (5 cycles should be complete in two minutes). Continue cycles of 30 compressions and two breaths until help arrives or if the patient shows some sign of life such as breathing or movement.

AED

If the second rescuer arrives (with an AED), he proceeds to switch on the AED, attach the defibrillator pads to the bare chest of the patient, placing one pad in the right infraclavicular region and the other on the precordium. He allows the AED to analyse the cardiac activity for a shockable rhythm. During analysis, the rescuers should not touch the patient. If the AED confirms the presence of a shockable rhythm, it automatically charges the pads and requests the user to press the shock button so that the shock can be delivered to the patient. The rescuer ensures that nobody is touching the patient or the bed, including himself, announces loudly and delivers the shock. The second rescuer takes over chest compressions whereas the first rescuer (who may be tired by then) would take care of airway and breathing. The two rescuers give chest compressions and ventilations at a ratio of 30 : 2 (Fig 48.34).

Rescuer fatigue can affect the quality of CPR. After five cycles of CPR (2 min), the rescuer must switch roles to provide compression and ventilation. The heart rhythm is analyse every two min to see whether there is a return of spontaneous cardiac activity. If the AED declares that there is no shockable rhythm, carotid pulse should be felt. If a palpable pulse is present, stop chest compressions and continue rescue breathing as necessary

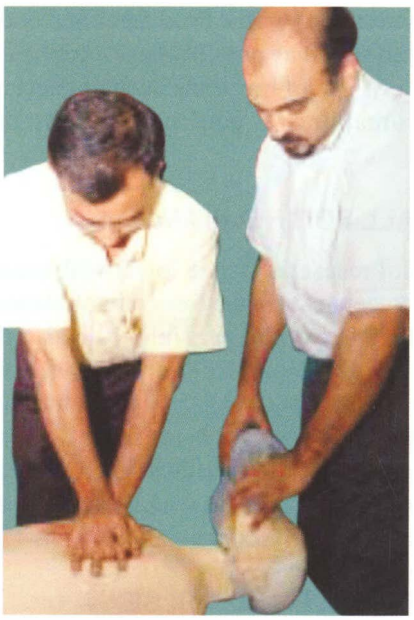


Fig. 48.34: Two rescuer CPR

(1 breath every 5 seconds). If breathing also returns and is adequate, the patient may be allowed to breathe spontaneously. The victim may be turned to a lateral position (recovery position) and monitored. The victim must then be shifted to a hospital for further evaluation and care.

BLS is usually continued till help arrives and advanced life support measures can be attempted. However, if no help is at hand, and if the rescuer is unable to continue due to fatigue or if the victim is obviously dead, CPR may be stopped.

Advanced CPR (advanced cardiac life support – ACLS)

Advanced CPR involves using airway adjuncts, other equipment and drugs for resuscitation.

A. Airway

Bag-mask ventilation (Fig 48.35), if effective in achieving chest expansion is adequate in the initial phase of CPR. Airway is best maintained with endotracheal intubation. However, chest compressions should not be interrupted to facilitate endotracheal intubation. If it can be achieved without interrupting chest compressions, then one may proceed to secure the airway with an endotracheal tube. The route of choice for intubation is through the mouth because it is faster and less traumatic.

Endotracheal intubation has the following advantages:

1. The trachea is protected from aspiration of regurgitated gastric contents.
2. The tidal volume can be assured
3. There is no distention of the stomach.

In case endotracheal intubation is difficult either due to difficult airway or inexperience in the technique, oxygenation and ventilation can be achieved by the use of an oropharyngeal

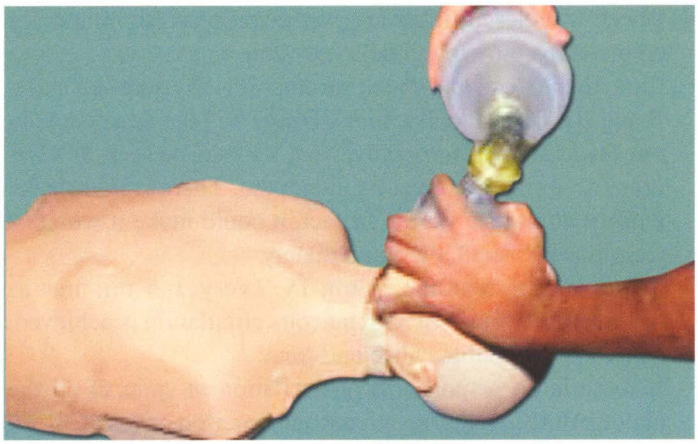


Fig. 48.35: Bag-mask ventilation

or nasopharyngeal airway along with a self-inflating bag and face mask. Usually this is adequate but occasionally a laryngeal mask airway or a Combitube® along with a self-inflating bag may also be used.

B. Breathing

After the airway is secured, **ventilation** must be provided at a rate of **8–10 breaths per minute** using a self-inflating bag attached to the endotracheal tube. **Oxygen** must be attached to the bag to improve oxygenation. These breaths need not be interposed between cycles of 30 compressions. Once the airway is secured with an endotracheal tube, chest compressions are given uninterrupted at a rate of at least 100/min while breaths are given every 6–8 seconds. It is important not to hyperventilate during CPR as it can produce constriction of cerebral arteries, reduce cerebral blood flow and adversely affect outcome.

A capnograph may be attached to the endotracheal tube to monitor end-tidal carbon dioxide. **A capnograph has three uses in CPR:** 1) To confirm the placement of endotracheal tube 2) To monitor the effectiveness of CPR. If the end-tidal carbon dioxide concentration is >10 mm Hg, the cardiac compressions can be deemed effective and 3) A sudden increase in end-tidal carbon dioxide to normal or near normal levels heralds a return of spontaneous circulation (ROSC). Thus, when an endotracheal tube is in place, efforts must be made to obtain and use a capnograph, whenever possible.

C. Circulation

Continue cardiac compressions as in BLS. Obtain peripheral intravenous access quickly.

D. Drugs/Defibrillation

Cardiac arrest rhythms are one of the following three types: Ventricular fibrillation/Ventricular tachycardia (VF/VT), pulseless electrical activity (PEA) and asystole. A monitor is obtained as early as possible in CPR to diagnose these rhythms.

VENTRICULAR FIBRILLATION/VENTRICULAR TACHYCARDIA

Most of the adult nontraumatic cardiac arrests are due t ventricular fibrillation (VF)/ventricular tachycardia (VT). I untreated, VF/VT will degenerate into asystole. The onl treatment of ventricular fibrillation is electrical defibrillatior The success rate of defibrillation reduces by 10% every passin minute. Hence, defibrillator must be obtained as early a possible (Fig 48.36).

Fig. 48.36: A manual defibrillator

If the rhythm shows ventricular fibrillation (VF) or ventricular tachycardia (VT) (Figs 48.37 and 48.38), the following algorithm must be followed:

- Provide Basic CPR. Once VF/VT is confirmed, interrupt CPR briefly to deliver a single **shock** of 360 J (monophasic) or 200 J (biphasic). **Resume CPR** immediately and continue for another two minutes (5 cycles of compressions and ventilations) before reanalyzing the rhythm.

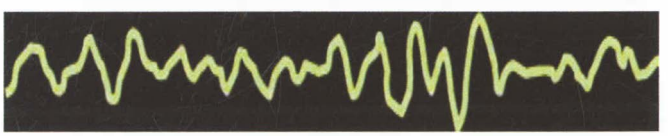


Fig. 48.37: ECG rhythm showing ventricular fibrillation. Note the saw tooth shaped waves that are irregular and chaotic in pattern

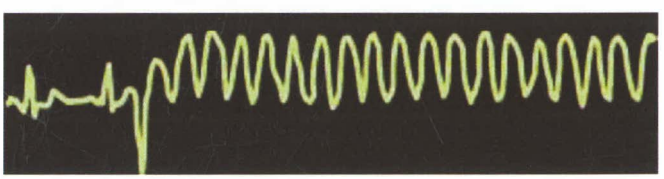


Fig. 48.38: ECG rhythm showing ventricular tachycardia which is more regular and shows wide QRS complex with inverted T waves

- If VF/VT persists, deliver another single **shock** of 360 J (monophasic) or 200 J (biphasic) and **resume CPR**. Administer **adrenaline** 1 mg **or vasopressin** 40 IU (one dose only). Reassess after two minutes.
- If VF/VT persists, deliver another single **shock** of 360 J (monophasic) or 200 J (biphasic) and resume CPR. An antiarrhythmic drug such as **Amiodarone** 300 mg may be given IV.
- Thus, analysis of the rhythm is repeated every two min and if a shockable rhythm is present, shock is given. Epinephrine 1 mg is repeated every 3-5 min. A second dose of amiodarone 150 mg may be considered for persistent VF/VT. Always remember to look for treatable causes (Key Box 48.6) of arrest.

PULSELESS ELECTRICAL ACTIVITY (PEA)

If the monitor showed no VF/VT, but a discernible rhythm, sinus or nonsinus with the patient manifesting no pulse (no carotid pulsations), it is termed pulseless electrical activity (PEA). This means that electrical activity is present while mechanical activity is absent.

While PEA may be caused by massive myocardial infarction where the myocardial necrosis is so severe that ventricular contractions are absent **(electromechanical dissociation - EMD),** it may also be caused by the following factors which may result in ineffective contractions while electrical activity is still present **(pseudo-EMD)**. In pseudo-EMD, the causes are reversible. If the cause is identified and treated promptly, the chances of successful resuscitation are high.

1. Commence Basic CPR
2. This must be followed by **adrenaline,** 1 mg IV, every 3–5 minutes as necessary till return of spontaneous circulation is achieved.
3. Look for a treatable cause (Key Box 48.6) and treat promptly.

KEY BOX 48.6

REVERSIBLE CAUSES OF CARDIAC ARREST

1. **H**ypoxia	1. **T**ension pneumothorax
2. **H**ydrogen ions (acidosis)	2. **T**amponade (Cardiac)
3. **H**ypovolaemia	3. **T**hrombosis (Pulmonary, Coronary)
4. **H**yper/hypokalaemia	4. **T**oxins
5. **H**ypo/hyperthermia	5. **T**rauma

Remember **5 Hs, 5 Ts**

ASYSTOLE

If the monitor shows a flat line, it is asystole. Whenever asystole is seen on a monitor the following things should be done (flat line protocol). Confirm that it is asystole and then commence therapy.

- Check whether all leads are connected correctly. Improper connections may manifest as asystole.
- Change the Lead and check whether it is asystole in Lead I, II and III. Occasionally, fine VF may be seen as asystole in one lead but on changing the lead, VF may be seen. If VF is seen, defibrillation must be attempted. There is no role for defibrillation in asystole. In fact, it could make it worse.
- Commence basic CPR immediately.
- Administer adrenaline, 1 mg IV, every 3–5 minutes as necessary till return of spontaneous circulation is achieved. Look for a treatable cause and treat.
- If there is no response to resuscitation in spite of all the above efforts even after a reasonable period of time (30–60 min) consider abandoning resuscitation efforts.

DRUGS USED IN CPR

1. Adrenaline

This is the drug of choice in cardiac arrest. It is given in a dose of 1 mg, diluted to 10 cc (1:10,000) intravenously every 3–5 minutes. This should be followed by at least 20 cc of 'chaser' solution (0.9% saline or Ringer lactate) and limb elevation for 15-20 s. A peripheral intravenous access is adequate but if a central line is in situ, the drug may be injected in the central line. If intravenous access is not available, an interosseous access may be secured (anteromedial aspect of tibia or lower end of femur may be used). Alternately, adrenaline may be injected through the endotracheal tube but higher doses need to be used (2–5 mg diluted to 10 cc).

Intracardiac administration of adrenaline is not advisable for the following reasons:

a. Possible injury to coronary arteries
b. Possible injury to the pleura
c. Accidental injection into the myocardium can precipitate resistant ventricular fibrillation
d. Since the right ventricle is anterior, intracardiac injection is usually into this chamber from which it has to traverse the pulmonary circulation to reach the coronary arteries.

This may well be achieved by an intravenous injection, running it in a drip and avoiding the complications of intracardiac injection. Intracardiac injection is advisable only when done under vision as during open heart surgery or thoracotomy.

2. Vasopressin

This is a potent vasoconstrictor that can be given as a single dose of 40 IU and as an alternative to the first or the second dose of adrenaline in a victim of cardiac arrest. The vasoconstriction produced raises the diastolic pressure and hence, the coronary perfusion pressure during CPR.

3. Amiodarone

If VF/pulseless VT persists in spite of the adrenaline/vasopressin followed by a DC shock of 360 J, amiodarone

may be given in a dose of 300 mg intravenously followed by 150 mg after five min, as necessary. Other drugs that may be used are: Lignocaine: 1–2 mg/kg intravenously or magnesium sulphate: 1–2 g intravenously. Not more than two anti-arrhythmic drugs may be used to treat an arrhythmia as all antiarrhythmics are also proarrhythmics and may also produce myocardial depression.

4. Sodium bicarbonate

An alkalinising agent is used in the following situations:

- When there is documented metabolic acidosis
- The cardiac arrest is due to hyperkalaemia or
- The arrest period is prolonged beyond 10 minutes.

The dose is 1–2 mmol/kg intravenously.

5. Calcium chloride/gluconate

Calcium is given only in the following situations:

- If the cardiac arrest is due to hyperkalaemia
- There is documented hypocalcaemia or
- The patient has been on calcium channel blockers.

The dose is 10 ml of 10% calcium chloride or gluconate. Thus, both calcium and sodium bicarbonate are indicated when there is hyperkalaemic cardiac arrest. Their routine use in cardiac arrest is not indicated as they can cause more harm.

POSTRESUSCITATION CARE

If return of spontaneous circulation is achieved, it is important to ensure it is sustained. Cardiac compressions are stopped when the pulses begin to be felt without the compressions. Intravenous fluids and/or an infusion of an inotrope such as adrenaline or dopamine may be required to sustain normal blood presasure. A blood sample may be taken to quickly analyse the cause of cardiac arrest such as acidosis, disturbances in potassium levels, poor oxygenation, and for cardiac enzymes. A 12-lead electrocardiogram may be obtained to rule out ST-elevation MI. If present, the patient will require cardiology intervention such as coronary revascularisation. The patient will need further care in the intensive care unit where respiratory and renal support can be provided. It is important to preserve cerebral function by maintaining normal blood pressures, normal blood gases and pH, and normal cerebral perfusion pressures. If the patient does not regain consciousness after CPR and ROSC, consider induced hypothermia by infusing one litre of cold saline (4° C) with a target body temperature of 32 to 34°C. This is done in an attempt to reduce cerebral metabolism so as to better preserve brain function while the patient recovers from the after-effects of cardiac arrest and CPR.

Thus, cardiopulmonary resuscitation should be aimed at recovery of all functions of the body including the brain and is more aptly called cardiopulmonary cerebral resuscitation (CPCR).

PRECARDIAC ARREST RHYTHMS

Often cardiac arrest follows a period characterized by som abnormal rhythms such as symptomatic bradycardia (heart ra < 50/min or symptomatic tachycardia (heart rate > 150/min The symptoms are usually chest pain, dizziness and syncop palpitations or generally feeling weak. If these are identifie and treated promptly, cardiac arrest may be averted. Oxyge must be administered. Attach a monitor and secure a intravenous access.

Symptomatic bradycardia

Symptomatic bradycardia may be due to sinus bradycardia c heart blocks. Atropine (boluses of 0.5 mg IV, repeated to maximum of 3 mg) is the drug of choice followed by a infusion of adrenaline (2–10 mg/min) or dopamine (2–10 mg kg/min). If the bradycardia is unresponsive to atropine dopamine or adrenaline, a transcutaneous or transvenou pacemaker may be required to pace the heart. Always look fo and treat the cause.

Symptomatic tachycardia

Cardiac arrest may also be preceded by symptomati tachycardia (heart rate > 150/min). The symptoms are due t inadequate circulation or the tachycardia itself such as ches pain, dizziness and syncope, palpitations or generally feelin weak.

Symptomatic tachycardia may be due to sinus tachycardia This may be due to causes such as pain, fever, sepsis, fear hypovolaemia, etc. Identify and treat the cause.

- If the patient is having symptomatic tachycardia, which i not sinus in origin, measure blood pressure. If it is low (<90/60 mm Hg) and the patient is unstable, proceed t synchronised cardioversion (shock levels will depend or the rhythm – supraventricular tachycardia and monomorphic ventricular tachycardia – 100 J, atrial fibrillation—200 and polymorphic ventricular tachycardia – 360 J – all shock strengths given for monophasic defibrillator).
- If the patient is stable and has supraventricular tachycardia the options are vagal manoeuvres (carotid sinus massage or Valsalva manoeuvre), adenosine (6 mg rapid IV push followed by 12 mg × 2 doses as necessary) or simply rate control with beta blockers or calcium channel blockers.
- If the patient is stable and has broad QRS complex tachycardia, the options are adenosine (6 mg rapid IV push followed by 12 mg × 2 doses as necessary) to rule out supraventricular tachycardia with aberrant conduction. If no response or ventricular tachycardia is suspected, administer 150 mg of amiodarone, diluted in 5% dextrose and given IV over 10 min. If no response, can consider repeating the dose.
- Consider expert consultation from cardiologist at all times.

CLINICAL NOTES

HE LIVED TO TELL HIS TALE!

45-year-old doctor had a cardiac arrest due to myocardial infarction. He was given effective CPR promptly. In addition since the aetiology was ischaemic heart disease, he was taken to cardiac catheterisation laboratory where his myocardium was revascularised (PTCA). He received CPR all through this procedure. His heart had stopped for 45 minutes but started beating once revascularisation was complete. He required intensive care for cardiorespiratory support initially and for complications such as renal failure later. He went on to make a full recovery after a few weeks.

His recovery was so complete that he could present his own case in a conference later! This case is being mentioned here only to illustrate the following:

- *Although survival rates are low, it is still possible to save lives with a timely and well-performed CPR.*
- *CPR is only a symptomatic therapy. Treating the cause of arrest is equally important.*
- *Good postresuscitation care is essential to ensure recovery of all body systems.*
- *It is possible to recover good cognitive function with good CPR. So, it is better termed cardiopulmonary cerebral resuscitation (CPCR).*

PEARLS OF WISDOM

Students should aim to be able to quickly identify various life-threatening and nonlife-threatening arrhythmias and act accordingly. Students are urged to refer medical textbooks for features of various nonlife-threatening arrhythmias. However, always treat the patient and not the monitor!

PEARLS OF WISDOM

The outcome of cardiopulmonary resuscitation depends on the time of commencement as well as quality of CPR. The author feels that certification in basic as well as advanced CPR must be made mandatory for all doctors.

PEARLS OF WISDOM

Delivery of good quality CPR cannot be learnt from books. The students are urged to get their certification from a recognised training facility.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- Current CPR guidelines have been given with emphasis on cardiac compressions.
- Role of capnography in CPR has been added.
- Section on postcardiac arrest care is more elaborate.
- Importance of identification and treatment of precardiac arrest rhythms has been added.

MULTIPLE CHOICE QUESTIONS

1. With the mouth wide open and tongue protruded in a patient in sitting position, if only soft and hard palate are seen, his airway is classified as Mallampati class:

A. I
B. II
C. III
D. IV

2. On direct laryngoscopy, if only the epiglottis is seen, the view is said to be Cormack and Lehane:

A. I
B. II
C. III
D. IV

3. The following period is adequate fasting before administration of anaesthesia after taking a glass of cow's milk:

A. 2 hours
B. 4 hours
C. 6 hours
D. 8 hours

4. American society of anaesthesiologists physical status (ASA PS) 6 would be appropriate to describe the following patient

A. Patient with moderately controlled diabetes mellitus on insulin
B. Patient posted for surgery for 6 times before
C. Patient in severe hypovolaemic shock undergoing fluid resuscitation
D. Brain dead patient for organ donation

5. The following drug is *not* helpful in reducing the effects of aspiration:

A. Sucralfate
B. Metoclopramide
C. Pantoprazole
D. Atropine

6. Adrenaline is given in cardiopulmonary resuscitation because:

A. It stimulates the heart
B. It counters the effects of histamine
C. It is a bronchodilator
D. It is a powerful vasoconstrictor

7. The following is an ideal anaesthetic agent for inhalation induction:
A. Isoflurane
B. Sevoflurane
C. Desflurane
D. Diethyl ether

8. Succinylcholine is contraindicated in all of the following *except*:
A. Full stomach
B. Raised intracranial pressure
C. Open eye injury
D. Crush injury

9. The following inhalation anaesthetic requires a heated vaporiser:
A. Isoflurane B. Sevoflurane
C. Desflurane D. Diethyl ether

10. The following anaesthetic can be given by nasal, intramuscular and intravenous routes for induction of anaesthesia:
A. Thiopentone B. Ketamine
C. Etomidate D. Propofol

11. The following intravenous anaesthetic is useful in status asthmaticus for its bronchodilatory effect:
A. Thiopentone B. Ketamine
C. Etomidate D. Propofol

12. The following intravenous anaesthetic produces dissociative anaesthesia:
A. Thiopentone
B. Ketamine
C. Etomidate
D. Propofol

13. The following local anaesthetic is also Class 1b antiarrhythmic agent:
A. Lignocaine B. Bupivacaine
C. Prilocaine D. Cocaine

14. The following local anaesthetic is very cardiotoxic:
A. Lignocaine B. Bupivacaine
C. Prilocaine D. Ropivacaine

15. The following anaesthetic is described as an antanalgesic:
A. Thiopentone B. Ketamine
C. Etomidate D. Propofol

16. The compression-ventilation ratio in Basic CPR in an adult is:
A. 30:2 B. 15:2
C. 5:1 D. 3:1

17. The following drug is absolutely contraindicated in acute intermittent porphyria:
A. Thiopentone
B. Ketamine
C. Etomidate
D. Propofol

18. The following muscle relaxant produces 'accommodation blockade':
A. Succinylcholine
B. Atracurium
C. Vecuronium
D. Tubocurarine

19. Neostigmine is used in all of the following situations *except*:
A. Myasthaenia gravis
B. Reversal of nondepolarising muscle relaxant effect
C. Cobra bite
D. Cholinergic crisis

20. The gold standard for confirmation of endotracheal tube position is
A. Presence of bilateral air entry
B. Absent breath sounds in the epigastrium
C. A square wave capnogram
D. Bilateral visible chest rise

ANSWERS

1 C	2 C	3 C	4 D	5 D	6 D	7 B	8 A	9 C	10 B
11 B	12 B	13 A	14 B	15 A	16 A	17 A	18 A	19 D	20 C

49

Organ Transplantation

Principles of Transplantation
- Pathophysiology
- Major histocompatibility complex
- Graft rejection

Liver Transplantation
- Indications
- Contraindications
- Donor criteria
- MELD score
- Types
- Procedures

Renal Transplantation
- Donor
- Procedures
- Postoperative management
- Complications

Small Bowel Transplant
- Indications
- Contraindications
- Recepient evaluation
- Procedures
- Complications

Islet Cell Transplantation
- Indications
- Contraindications
- Islet cell preparation
- Technique
- Complications

Drgus used for Immunosuppression

PRINCIPLES OF TRANSPLANTATION

Introduction

For end stage renal or liver disease, transplantation is the best answer as of today. Over 25,000 transplantations are performed annually for different conditions and more than 100,000 patients are awaiting an organ for kidney, liver or other organs. The acceptance of an organ depends often on the beliefs and decisions related to the country, race and religion. In certain countries, living donor transplantation, even though risky, is the only realistic organ donation method, greatly limiting the potential for transplantation of organs from cadaver. Surgical advances have permitted successful transplantation of lung, pancreas and intestine from living donors. Because of the limited supply; use of living donors may be the only means to achieve timely kidney or liver transplantation as it is happening in India. First let us study the principles and pathophysiology of organ transplant.

Pathophysiology of organ transplant

Generally the word "immunity" is used in the context of infections. "Immunity" implies defense against infections. Immunity is of two types, namely:

1. Innate immunity
2. Adaptive or acquired immunity

Innate immunity: Important components of innate immunity are:

- Phagocytic neutrophils
- Natural killer cells (NK cells)
- Circulating plasma proteins mainly complements.

Adaptive immunity: Adaptive immunity is normally quiescent but is pressed into action by the presence of suitable stimulus such as microbes. Components of adaptive immunity are lymphocytes and their products. Adaptive immunity may be

- Humoral immunity—mediated by antibodies which are synthesised by B-lymphocytes (B cells). They offer protection against extracellular microbes.
- Cell mediated immunity—mediated by T-lymphocytes (T cells). T cells may be CD4 T cells which indirectly facilitate killing of microbes by macrophages or CD8 T cells which directly kill microbes. CD8 T cells are also called cytotoxic T cells.

Major histocompatibility complex (MHC)

The MHC complex is also known as human leucocyte antigen (HLA) complex. It consists of a cluster of genes located on chromosome 6. MHC gene products are displayed on the cell surface to the notice of circulating T cells. Such MHC gene products fall into two categories.

1. Class I MHC molecules—which display peptides synthesised within the cytoplasm of the respective cell. Class I MHC molecules are present in all cells.

2. Class 2 MHC molecules—which display antigens synthesised outside the cell. Class 2 MHC molecules are mainly expressed in antigen processing cells (APCs) or dendritic cells.

Role of MHC in organ transplant

Following an allograft, the host cells recognise the foreign nature of the graft by two mechanisms.

1. Direct recognition—the T cells recognise the foreign class 1 MHC because of immunologic cross reaction. Subsequently cytotoxic T cells are activated which kill the graft cells.
2. Indirect recognition—the host APCs present the antigens in the graft (transplant) in class 2 MHC. This activates T cells which secrete cytokines, induce inflammation and damage the graft.

Towards improving graft survival

- ABO blood group antigens are expressed in all cells and are not just by RBCs. ABO incompatibility results in hyperacute rejection of graft. Hence, it is recommended to ensure ABO compatibility.
- Better HLA matching improves transplant outcomes. This is particularly relevant in live donor kidney transplant. However, HLA matching is not done in heart, lung and liver transplants as the urgency of transplant requirement overweighs the benefits of HLA matching. Also in these situations other factors such as size of the graft assume more practical significance.

Graft rejection

Based on the timing and mediators of rejection, graft rejection may be classified as follows:

1. **Hyperacute rejection**
   - Occurs within minutes to days following transplant
   - Mediated by preformed antibodies
   - Untreatable but preventable
   - Cross-matching prevents hyperacute rejection
   - Methods of cross-matching include—lymphocytotoxic assay, flow cytometric technique, bead-based screening assays, panel-reactive antibody assay.
2. **Acute rejection**
   - Occurs in weeks to months following transplant
   - Mediated by T cells to a great extent and B cells to a lesser extent.
   - Accordingly they are termed T cell-mediated rejection (TCMR) or antibody mediated rejection (ABMR).
   - Immune suppression helps to overcome acute rejection
3. **Chronic rejection**
   - Most common cause of long-term allograft loss
   - Occurs over months to years following transplant
   - Involves both T cells and B cells
   - Examples for chronic graft rejection—interstitial fibrosis, tubular atrophy and chronic allograft nephropathy in renal transplant, vanishing bile duct syndrome in liver transplant and bronchiolitis obliterans in lung transplant.

LIVER TRANSPLANTATION

Introduction

Liver transplantation is a lifesaving procedure for patients who have chronic end-stage liver disease and acute liver failure (ALF) when there are no alternative treatment options. In 1963 Dr Thomas Starzl performed the first three human liver transplantations at the University of Colorado but patients suffered from biliary atresia, had coagulopathy and did not survive surgery. Introduction to cyclosporine for immunosuppression in solid organ transplantation revolutionised the liver transplantation surgery.

Indications for liver transplant

1. **Fulminant hepatic failure**: It is acute onset of liver failure in the absence of pre-existing liver disease. Here coagulopathy sets in within 8 weeks of onset of jaundice. Causes are acetaminophen overdose and hepatitis B infection in Asia. If left untreated, patients most often succumb to coma due to cerebral oedema.
2. **Hepatitis C**: Chronic hepatitis C is the commonest indication for liver transplant in the West. However, hepatitis C recurs even after liver transplantation because the virus persists in the extrahepatic tissues. Pre-transplant and post-transplant treatment with interferon and ribavirin can potentially improve the outcome in these patients.
3. Hepatitis B
4. Primary biliary cirrhosis
5. Primary sclerosing cholangitis
6. Alcoholic liver disease
7. Nonalcoholic steatohepatitis (NASH)
8. Biliary atresia—it is the commonest indication for liver transplant in the paediatric age group.
9. **Hepatocellular carcinoma:** Theoretically liver transplant should offer the best chance of cure in HCC but there is always the risk of recurrence of HCC following transplant. Many transplant centres follow "Milan criteria" to decide the feasibility of liver transplant in HCC, which says that liver transplant can be considered in HCC when—there is a single nodule of < 5 cm or there are fewer than 3 nodules, the largest of which is measuring < 3 cm. Those who follow the "**Milan criteria**" believe that the recurrence of HCC is very low if the criteria are strictly adhered to.

Contraindications for liver transplant

1. Systemic infections other than liver infections
2. Pulmonary manifestations of chronic liver disease
3. Inability to abstain from alcohol when liver transplantation is being contemplated for alcoholic liver disease.
4. Lack of commitment to immunosuppressive drugs
5. Metastatic HCC

Following are **not contraindications** for liver transplantation:

- Failure of other organs in addition to liver, e.g. amyloidosis—here combined liver and kidney transplants can be undertaken.

- HIV infection
- Portal vein thrombosis

Donor criteria for liver transplantation: Donors for liver transplant can be deceased donors or live donors. The outcomes are better with deceased donors, but the limitation is shortage of such donors. The ideal donor should be young and otherwise healthy. The donor factors associated with increased risk are, older age, a fatty infiltration and use of split liver from these donors. Two categories of donors who are increasingly considered although not ideal are:

- **Older donors:** If liver from older donors are used for hepatitis C recipients, there is an increased risk of development of cirrhosis in the transplanted liver.
- **Donor after cardiac death:** These are donors who do not meet brain death criteria but become donors once life support is taken off. Liver from such donors can be considered for transplant with cut off time of 30 minutes.

Model for end-stage liver disease (MELD score)

Candidates needing liver transplant far outnumber the available donors. Therefore candidates needing liver transplant are listed in the transplant register in organ transplant programmes. The order in which candidates are placed in the waiting list for liver transplant is determined by MELD formula. MELD formula incorporates three components, namely—creatinine, bilirubin and INR. These components provide an objective assessment of severity of liver disease.

MELD score =

$$[0.957 \times \text{Ln creatinine (mg/dl)}] + [0.378 \times \text{Ln bilirubin (mg/dl)}] + [1.120 \times \text{Ln INR}]$$

In the waiting list, candidates with higher MELD score come first. MELD score is nowadays preferred to Child-Pugh's score for being more objective.

Types of liver transplantation

1. **Conventional liver transplant:** This is performed to replace the diseased liver with a healthy liver from a deceased donor.
2. **Expanded criteria donor (ECD):** A diseased donor over the age of 60 with mild liver abnormalities. The term "expanded" is used because an expansion of the donor pool is considered to increase transplantation. With an ECD liver, the waiting time may be shorter—not done in India.
3. **Living donor liver transplantation (LDLT):** A procedure in which a healthy, living person donates a portion of his or her liver to another person. Finding a living donor match shortens waiting time, increases long-term transplant success and gives the flexibility of scheduling the date of surgery.
4. **Split liver transplant:** A deceased donor liver is split into two functioning units, which are used for transplantation to a child (left lobe) and an adult (right lobe).
5. **Combined organ transplant:** A person may receive more than one organ during the same transplant procedure such as liver and kidney or liver and heart. This may be recommended for patients who are experiencing multiple organ failure.

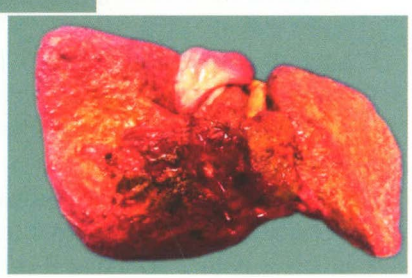
Technical aspects of liver transplantation

More often the donor in liver transplant is deceased. However, with severe shortage of donors, live donor liver transplants are becoming common. The donor liver transplant is placed in its native position in the abdomen of recipient after removing the diseased liver. Hence, the term "Orthotopic liver transplant" is used. (This may be compared with kidney transplant where the transplanted kidney is placed in the iliac fossa thereby deriving the term heterotopic transplant.) Sometimes, the donor liver is split and the left liver is used for a paediatric recipient and the right liver is used for an adult recipient. Such a liver transplant is termed "Split liver transplant". A few operative steps in the back table where the donor live is prepared for transplantation) is given in Key Box 49.1.

KEY BOX 49.1

KEY POINTS IN THE BACK TABLE SURGERY

1. Dissection and removal of extra tissue such as diaphragm, adrenal gland, pancreatic, tissue etc.
2. Preparation of cuffs of the suprahepatic and infrahepatic vena cava, cleaning of the portal vein and artery, and inspection of the bile duct.
3. Verification of secure ligatures on small retrohepatic caval, portal vein and hepatic arterial branches.
4. Confirm the continuity and integrity of all major structures that must be anastomosed to the companion recipient structures.



Figs 49.1 and 49.2: Cirrhotic liver which was removed (*Courtesy:* Dr Sachidananda N, hepatobiliary and liver transplant surgeon, Prof of Surgery, Akash Institute of medical Sciences and Research, Devanahalli, Bangalore)

Immunosuppression after liver transplantation

Chronic rejection is uncommon following liver transplantation. The need for immunosuppression decreases over time. Combination of calcineurin inhibitors (tacrolimus, cyclosporin), steroids (methylprednisolone) and antiproliferative agents (mycophenolate mofetil) are used. In the context of liver transplantation sirolimus deserves a special mention:

- It has antineoplastic activity, hence, is an attractive option in HCC.
- It delays wound healing
- It is associated with hepatic artery thrombosis

Complications of liver transplantation

1. Primary non-function—these patients need retransplant
2. Hepatic artery thrombosis—these patients need early thrombectomy or retransplant.
3. Portal vein thrombosis
4. Bile duct leak and stricture
5. Other complications—bleeding, coagulopathy

Future trends

Hepatocyte transplantation: In chronic liver diseases due to specific enzyme deficiencies like Crigler-Najjar syndrome, hepatocytes or stem cell transplant seems logical. With only a few cells needed to restore functioning of liver, this concept makes sense as it avoids the morbidity and mortality of a major surgical procedure.

RENAL TRANSPLANTATION

Preparation for transplantation

Recipient is prepared by haemodialysis. It also enhances the chances of success of a transplant.

Donors: Two sources

A. Cadaver kidney is obtained from 'brain dead' patients who are still living with mechanical ventilatory support. Consent should be taken from patient's relatives.

B. Living
   Living related
   Living non-related

Criteria of an ideal donor

1. Less than 60 years of age
2. No previous renal disease
3. No diabetes
4. No hypertension
5. Adequate renal perfusion 50 ml/hour urine output is necessary.
6. No systemic infections such as hepatitis B, C, HIV infection.
7. No malignancy

Living related donors

- An identical twin is ideal
- Father or mother
- Son or daughter
- Brother or sister

Tests done before transplantation

- Blood group ABO compatibility
- Biochemical investigations
- Complete blood picture
- Renal function tests
- Rule out: Diabetes, hypertension, hepatitis B infection hepatitis C infection, HIV.
- Bilateral renal angiography to study vascular pattern. Tissu typing.

PEARLS OF WISDOM

Recipient contraindication to renal transplantation is if the recipient is suffering from diseases such as heart disease or malignancy (advanced), which compromises his survival.

Operation technique

Donor operation

1. **Living related donor:** Donor nephrectomy witl preservation of as much length of the artery, vein and ureter
2. **Cadaver donor:** The Donor kidney is perfused with icy perfusion fluid and removed along with vena cava and aorta and packed in plastic bags surrounded by ice in an insulatec box. It can be stored like this for 72 hours.

Perfusion fluid: It has high concentration of potassium (8(mmol/L) and high osmolarity (400 mOsm/kg). This is usec during transplant anastomosis.

Recipient operation: Kidney is transplanted in the righ iliac fossa by anastomosing renal artery to internal iliac artery and renal vein to external iliac vein. Ureter is implanted in the bladder (Figs 49.3 to 49.5).

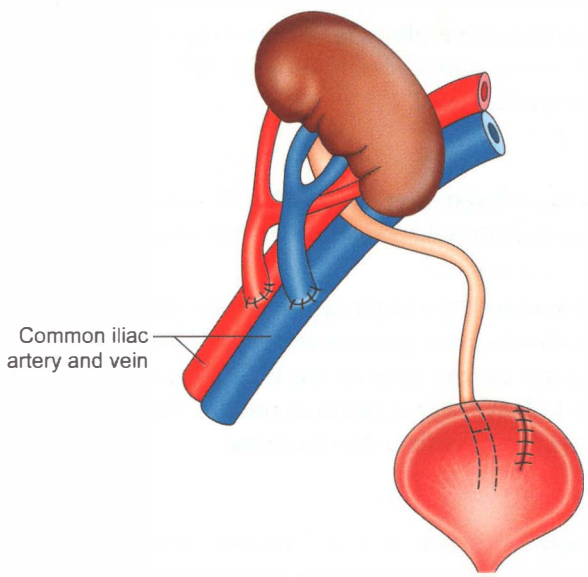
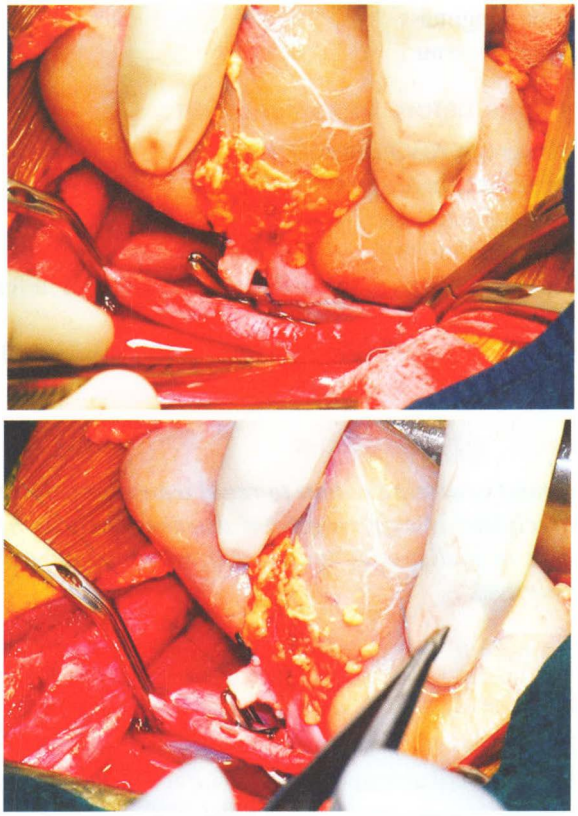


Fig. 49.3: Diagrammatic representation of renal transplantation



Figs 49.4 and 49.5: Renal artery and renal vein are ready to be sutured to common iliac artery and vein during renal transplantation

Postoperative management

1. **Immunosuppression:** Cyclosporin A is given alone or in combination with low doses of azathioprine and steroids.
2. **Fluid balance:** Aim to maintain urinary output at the rate of 20 ml/hour. Even an anephric patient needs 500 ml of fluid/day to replace insensible loss. CVP is ideal for management of fluids in the postoperative period.
3. **Postoperative oliguria:** Monitoring of renal function—Serum creatinine should drop by 50% in 48 hours post-transplant period.

Causes

a. Acute tubular necrosis (ATN): Minimal ATN is common due to short ischaemic time. Hence, oliguria phase may be a few hours.

b. Rejection: More likely on the 5th day. Twice weekly DTPA (diethyl triamine penta-acetic acid) isotope renography and gamma camera scan of the kidney confirms perfusion of the kidney when oliguria is present. Percutaneous needle biopsy can confirm the rejection.

Treatment of rejection

1. **Acute rejection:** Within 3 months, this responds to high doses of intravenous steroids. 90 mg methylprednisolone IV/day × 3 days.
2. Chronic rejection involves the vascular element. It does not respond to steroids.

Surgical complications

1. **Haemorrhage** manifests as oliguria, hypotension. Wound should be re-explored and bleeding vessels should be ligated.
2. **Renal artery thrombosis** results in nonfunctioning of the transplanted kidney. It can be diagnosed by ultrasonogram and isotope studies. This is treated by nephrectomy.
3. **Renal artery stenosis** can develop later. It should be treated by an angioplasty.
4. **Lymphocoele** can develop due to perivascular dissection and increased flow of lymph. Such lymphocoele can lead to ureteric obstruction. Small collection needs to be observed, large ones need drainage.
5. **Ureteric obstruction** or urinary leakage due to disruption of ureteroneocystostomy can be due to ischaemia or necrosis of the distal ureter. During donor nephrocystostomy, care should be taken not to remove too much of periureteric tissues and avoid too much dissection in the renal hilum to prevent ureteric ischaemia.

SMALL BOWEL TRANSPLANT

Introduction

- Small bowel is rich in lymphoid tissue and the large mucosal surface area with major histocompatibility antigens. This definitely evokes a fight between graft and host.
- Small bowel is colonised by a multitude of bacteria and other micro-organisms. This is one of the reasons for failure.

Indications

- The indication for intestinal transplant is intestinal failure which means inability to maintain protein-energy, fluid, electrolyte, or micronutrient balance. Most commonly it is due to the result of extensive small bowel resections following mesenteric ischaemia or following necrotising enterocolitis.
- **In children:** Children with massive intestine resection, children with severely diseased intestine and unacceptable morbidity, microvillus inclusion disease or intestinal epithelial dysplasia.

Contraindications

1. In individuals who have significant coexistent medical conditions that have no potential for improvement following transplantation.
2. If the patient has an active, uncontrolled infection
3. Malignancy

Pre-transplant recipient evaluation

- Complete blood picture
- Renal and liver function test

- Electrocardiogram.
- Rule out hepatitis, liver biopsy
- Upper GI scopy, colonoscopy
- Imaging—chest X-ray, barium studies
- USG abdomen and CT scan

Types and operative procedure

Three primary procedures

1. Isolated transplantation of the small intestine (with or without the portion of the right colon with the ileocaecal valve).
2. Combined transplantation of the intestine and liver as separate grafts.
3. Multivisceral transplantation (MVT), which is the simultaneous transplantation of a composite graft including the liver, stomach, duodenum, pancreas, and small intestine.

Details

1. Isolated transplantation of the small intestine: A few steps are given below

- A small bowel transplant from a deceased donor comprises the entire small bowel.
- The superior mesenteric artery of the graft (with an aortic patch) is anastomosed to the recipient aorta, and the superior mesenteric vein is anastomosed to the inferior vena cava or to the side of the portal vein.
- The proximal end of the small bowel graft is anastomosed to the recipient jejunum or duodenum.
- The distal end of the graft is anastomosed to the side of the colon.
- Protective loop ileostomy is done. It is an end-ileostomy.
- A gastrostomy tube is used for aspiration rather than to overcome delayed gastric emptying.
- Feeding jejunostomy tube is inserted.

2. Combined transplantation of the intestine and liver

- It is indicated when patients have cholestatic jaundice also secondary to TPN and require combined liver and small bowel transplantation.
- When combined liver and small bowel transplantation is carried out, the two grafts are transplanted *en bloc*.
- The donor aorta is fashioned into a conduit including the superior mesenteric and coeliac arteries and anastomosed to the recipient aorta.
- The portal vein anastomosis is similar to that in liver transplantation.

3. Multivisceral transplantation (MVT)

- Multivisceral, also called 'cluster' transplants may be necessary in the case of large desmoid tumours.

Postoperative management

- Induction of immunosuppression with monoclonal (alemtuzumab, basiliximab, daclizumab) or polyclonal (thymoglobulin) antibody given intraoperatively or preoperatively.
- Immediately following surgery, maintenance immuno suppression therapy is initiated by oral administration o tacrolimus. Steroids are also included in the postoperativ immunosuppressive regimen.

Complications

A. Rejection

a. Acute cellular rejection: Acute cellular rejection usuall occurs within the first year post-transplantation but ca occur at any time. Clinically, it can present as diarrhoea unexplained fever, abdominal pain and/or cramping.

b. Chronic rejection: Clinically, these patients may hav chronic diarrhoea despite adequate treatment. Patient have an obliterative arteriopathy.

B. Infection

1. **Bacterial:** Bacterial infections can manifest as intra abdominal infection, opportunistic infections, surgica site infections, pneumonia. Pathogenic organisms are *Escherichia coli, Klebsiella, Enterobacter, Enterococci* and commonly, polymicrobial infections.
2. **Viral:** Cytomegalovirus (CMV) is a common pathoge post-intestinal transplantation, which often affects th allograft. Treatment with ganciclovir and/or CMV immunoglobulin and rarely, reduction in immuno suppression are methods that are used to minimise graf loss.
3. **EBV (Epstein-Barr virus)** also presents a unique challenge to intestinal transplant recipients because o the higher rates of post-transplantation lympho proliferative disorder (PTLD) when compared with othe solid organ transplant recipients.

C. GVHD: Graft-versus-host disease (GVHD) occurs when donor lymphoid cells begin to target recipient tissues, most notably the epithelial cells in the skin and intestine. It is relatively uncommon.

D. Other complications: Postoperative haemorrhage, biliary (if liver is also used) thrombosis of vessels resulting in graft loss, bowel anastomosis leak and wound infections.

ISLET CELL TRANSPLANTATION

Introduction

- Type 1 diabetes mellitus, also known as juvenile-onset or insulin-dependent diabetes, is a chronic polygenic autoimmune disorder that has a strong hereditary basis in the human leucocyte antigen system. It results from destruction of pancreatic beta cells in the islets of Langerhans. Beta cells constitute 28–75% of pancreatic islets.
- Percutaneous islet cell transplantation is a minimally invasive cellular replacement therapy. It avoids risk of hypoglycaemia which is one of the major problems of exogenous insulin.

Indications

- Type 1 Diabetes mellitus (DM) with disease presence for at least 5 years, absence of endogenous C-peptide secretion.
- Islet cell transplantation may be performed alone, in combination with renal transplantation, or following kidney transplantation. Example: Diabetic patients with imminent or established end-stage renal disease.

Contraindications

- Age less than 18 years or greater than 70 years
- DM duration less than 5 years
- Residual C-peptide secretion (i.e. stimulated C-peptide level: 0.5 ng/dL).
- Proliferative diabetic retinopathy, portal hypertension
- Active infection (including hepatitis C, hepatitis B, HIV and tuberculosis).

Islet cell preparation

- After cold perfusion of the abdominal organs, the pancreas, spleen and duodenum is removed *en bloc*. The portal vein is transected at the duodenal border and the common bile duct is transected close to the pancreatic border.
- The organ is packed in a triple-barrier bag with cold preservation solution and stored at 4°C for transportation. The maximum cold ischaemia time is 12 hours.
- The most commonly used preservation solution is University of Wisconsin solution.
- **Obtaining islet cells:** The initial step is infusion of collagenase through the main pancreatic duct; the collagenase is delivered with pressure monitoring aimed at duct distension with minimum leakage of the enzyme. Collagenase cannot digest thick interlobar fibrous tissue. This is achieved by the **Ricordi chamber**. The chamber includes five or six stainless steel balls, which provide mechanical fracture of the fibrous tissue. Thus, it provides effective digestion, dilution and collection of the digested tissue. The purity of the final cell suspension is analysed and the cells are washed and placed in culture media for 12–72 hours in an incubator at 22°–37°C. The isolated islets are then transplanted.

Technique of transplantation

- The portal venous system is used for islet cell transplantation.
- With ultrasound (US) and/or fluoroscopic guidance, a branch of portal vein is punctured with a 20–22-gauge needle *via* a percutaneous transhepatic approach.
- The percutaneous access is dilated to accept a 5–6-F catheter or vascular sheath, which is advanced into the main portal vein.
- Confirm the catheter placement using ultrasound.
- Systemic anticoagulation with heparin should be initiated-5000 units is routinely administered.

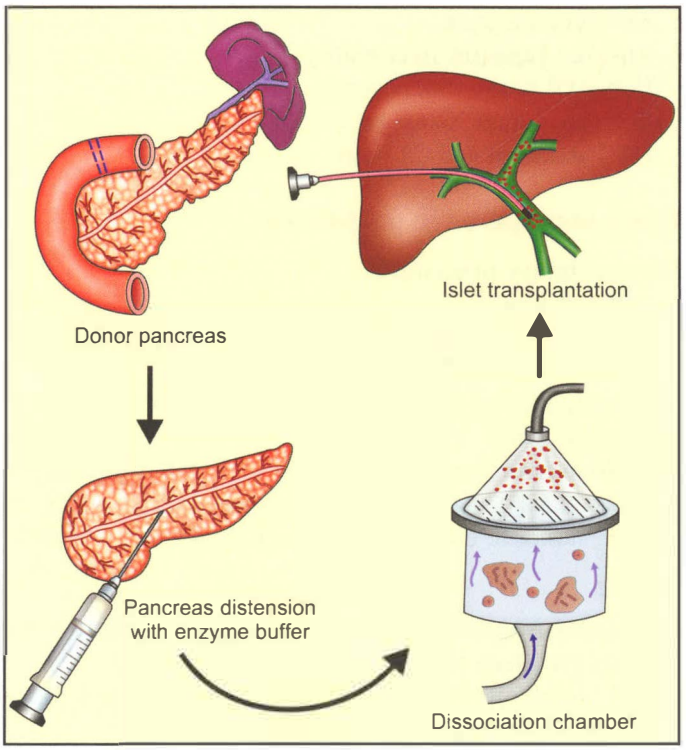
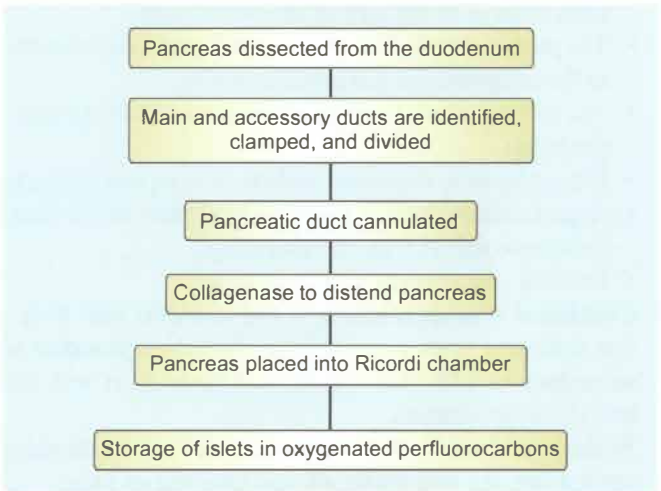


Fig. 49.6: Islet cells transplantation

Flowchart



Islet cell infusion

- At least 10,000 islet equivalents per kilogram of body weight.
- An islet equivalent refers to an islet measuring at least 150 µm in diameter. In general, harvested islet cells are infused by using gravity flow or direct syringe injection.

Peri- and post-procedural imaging, DM management, and immunosuppression

It includes a combination of a T cell-depleting agent and interleukin-2 monoclonal antibody receptor blocker (daclizumab) for induction and maintenance with a

combination of a calcineurin inhibitor (tacrolimus) and a mammalian target of rapamycin inhibitor (sirolimus).

Complications of islet cell transplantation

- Bleeding—either intraperitoneal or liver subcapsular is the most common procedure-related complication. Fortunately, effective hepatic parenchymal tract embolisation significantly reduces bleeding.
- Partial portal vein thrombosis is another complication. It is because of activation of coagulation system by transplanted cells.
- Other complications of islet cell transplantation include transient liver enzyme elevation, abdominal pain, focal hepatic steatosis and severe hypoglycaemia.

KEY BOX 49.2

OTHER SITES OF INJECTING ISLET CELLS

- Intrahepatic
- Renal subcapsular
- Intrasplenic
- Intraperitoneal
- Subcutaneous

Drugs used for maintenance therapy after transplantation

Steroids: Prednisolone is commonly used. Side effects ar hypertension, Cushing's syndrome, diabetes, cataract, muscl wasting.

Antiproliferating agents: Azathioprine is the drug used. I inhibits both humoral and cell mediated immunity. Bon marrow suppression and toxic hepatitis are side effects.

T cell directed immunosuppressants: Cyclosporine, Tacro limus and Sirolimus are a few drugs. Cyclosporine inhibit formation of mature CD4 and CD8 T cells in the thymus. I does not cause myelosuppression. Side effects are nephro toxicity, hypertension, hyperkalaemia, hirsutism, etc. It is use in a dose of 4 mg/kg in 500 ml saline, IV and later-oral therap with12 mg/kg daily. After a few weeks tapered to 5 mg/kg day.

Tacrolimus: Used in liver transplant and in acute rejection o the kidney. Effects are similar to cyclosporine. Side effect are also similar to cyclosporin but without hirsutism an gingival hypertrophy.

MULTIPLE CHOICE QUESTIONS

1. Which of the following is not a component of innate immunity?
A. Phagocytic neutrophils
B. Natural killer cells
C. Complements
D. Mast cells

2. Which of the following is a cytotoxic T cell?
A. CD8 B. CD4
C. CD6 D. CD2

3. Which are the following are included under MELD score in liver transplantation?
A. Creatinine, bilirubin, albumin
B. Creatinine, bilirubin, INR
C. Creatinine, bilirubin, bleeding time
D. Creatinine, bilirubin, ammonia

4. Tacrolimus is a:
A. Calcineurin inhibitor
B. Steroids
C. Antiproliferative agents
D. Antibacterial agent

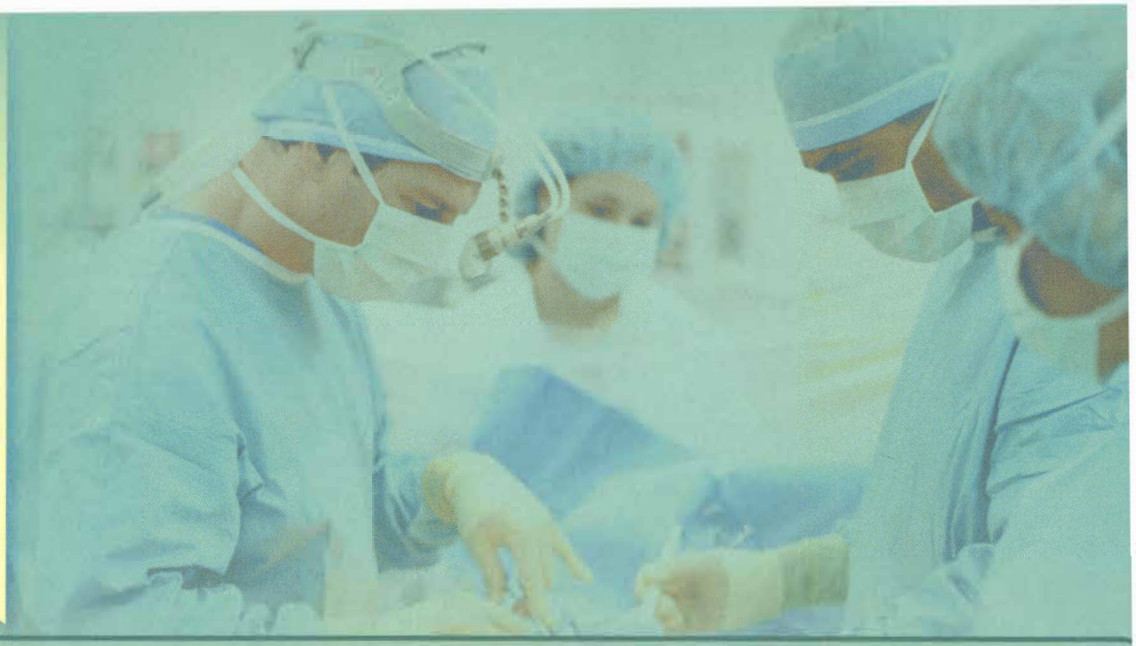
5. Following intestinal transplantation, which is the common intestinal pathogen causing infection?
A. Shigella
B. Cytomegalovirus
C. Streptococci
D. Meningococci

ANSWERS

1 C 2 A 3 B 4 A 5 B

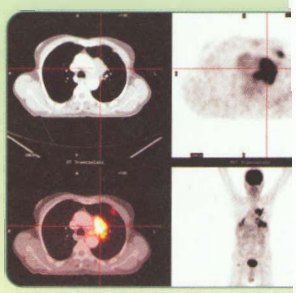
Section

V



Viva Voce Examination

50. X-rays and Images
51. Instruments
52. Specimens
53. Operative Surgery, Laparoscopic Surgery and Accessories



50

X-rays and Images

- Plain X-rays
- Barium swallow
- Barium meal
- Barium enema
- ERCP
- T-tube cholangiography
- Splenoportovenography (SPV)
- CT scan
- PET scan

Introduction

One of the sessions in the undergraduate and postgraduate examinations is on X-rays and images. More and more images such as ultrasound, CT scan, MRI and PET scan, etc. have been used. Hence I have added a few of them here. More details have been given in the chapter on radiology. Several CT images have been included in the book. Students are requested to study those CT scans and chapter on radiology first and then read this chapter. This is only an exercise for you to perform better in the final examination.

I. PLAIN X-RAY ABDOMEN SHOWING COLLECTION OF FREE GAS UNDER THE RIGHT DOME OF THE DIAPHRAGM

Normally, fundic air bubble is present on the left side. Hence, importance is given to the gas on the right side.

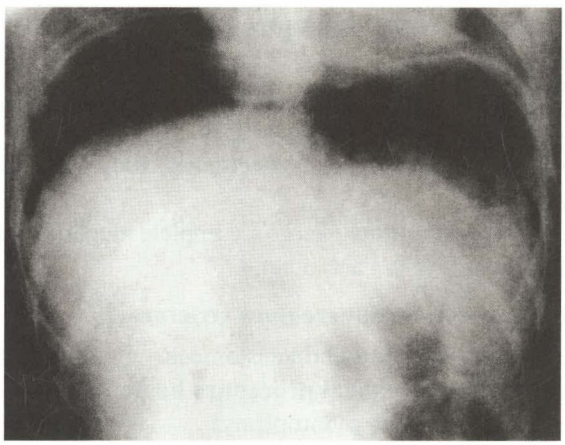


Fig. 50.1: Plain X-ray abdomen showing collection of free gas under the right dome of the diaphragm

1. What are the causes of free gas under the right dome of the diaphragm?

- Perforation of hollow viscus. Examples:
  - Duodenal ulcer, gastric ulcer
  - Perforation of enteric ulcer
  - Perforation of Meckel's diverticulum
  - Malignant ulcers—colonic, gastric
  - Perforation of the tuberculous ulcer—ileum
- Abdominal stab injury
- Laparotomy
- Tubal insufflation test done for tubal patency.

2. Is there any other finding in the X-ray?

- Ground glass appearance indicates significant fluid in the peritoneal cavity.

3. How do you manage a case of perforated duodenal ulcer?

- With antibiotic coverage, Ryle's tube aspiration and early resuscitation with intravenous fluids, exploratory laparotomy is done. The site of perforation is identified which is in the first part of the duodenum. The perforation is closed by using nonabsorbable sutures. Omentum can be used to reinforce the suture line. This is called Roscoe Graham operation. Corrugated red rubber drain or tube drain is used to drain peritoneal cavity.

4. Will you do elective surgery such as GJ and vagotomy or HSV at this stage?

- Since the general condition of the patient will be very poor at this acute stage because of hypovolaemic and septic shock, elective surgery is not done.

5. What are the stages of duodenal ulcer perforation?

- Stage of chemical peritonitis
- Stage of illusion or delusion
- Stage of bacterial peritonitis

II. PLAIN X-RAY ABDOMEN SHOWING MULTIPLE GAS AND FLUID LEVELS

1. What is the diagnosis?

- Since jejunal loops are prominently seen and loops are centrally located, it is probably terminal ileal obstruction.

2. What are the common causes of terminal ileal obstruction?

1. Tuberculous stricture
2. Bands—congenital
3. Adhesions
4. 'Worm ball' in children
5. Obstructed hernia

3. How do you identify jejunum, ileum and colon in a plain X-ray?

- Jejunum—valvulae conniventes—regularly placed mucosal folds placed opposite to each other.
- Ileum—no character—**characterless loop of Wangen-steen**.
- Colon—**haustrations**—large incomplete mucosal folds **not placed** opposite to each other.

4. How do you treat tuberculous strictures?

- Resection and end-to-end anastomosis

5. Can any other surgical procedure be done?

- Stricturoplasty (like pyloroplasty)

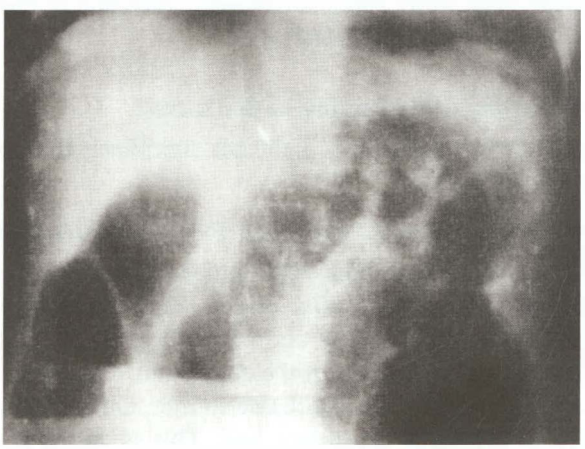


Fig. 50.2: Plain X-ray abdomen showing multiple gas and fluid levels

III. PLAIN X-RAY ABDOMEN SHOWING RADIO-OPAQUE SHADOW IN THE RIGHT UPPER ABDOMEN

1. What is the diagnosis?

- Probably renal stone

2. Why is it not a gall stone?

- The location of the stone is at lower level when compared to gall stone.
- The shape of the stone suggests that it is a stone in the pelvis growing within calyces.

3. What do you call such a stone?

- Staghorn calculus

4. What type of X-ray is ideal to distinguish renal stone from gall bladder stone?

- Lateral view

5. What will be the findings in case of renal stones in a lateral picture?

- Renal stones are found superimposed on vertebral bodies. On the other hand, gall stones are found anterior to it.

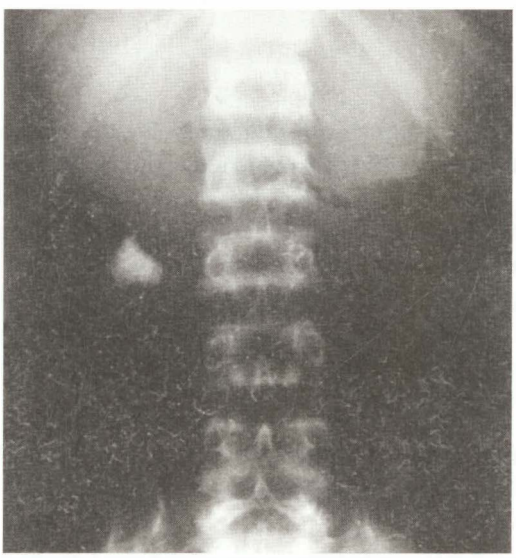


Fig. 50.3: Plain X-ray abdomen showing radio-opaque shadow in the right upper abdomen

IV. PLAIN X-RAY ABDOMEN SHOWING RADIO-OPAQUE SHADOW IN THE REGION OF GALL BLADDER

1. What is the diagnosis?

- Probably gall stone

2. What percentage of gall stones are visible in a plain X-ray?

- Only 10%

3. What is the reason for that?

- The calcium content in gall stones is very less.

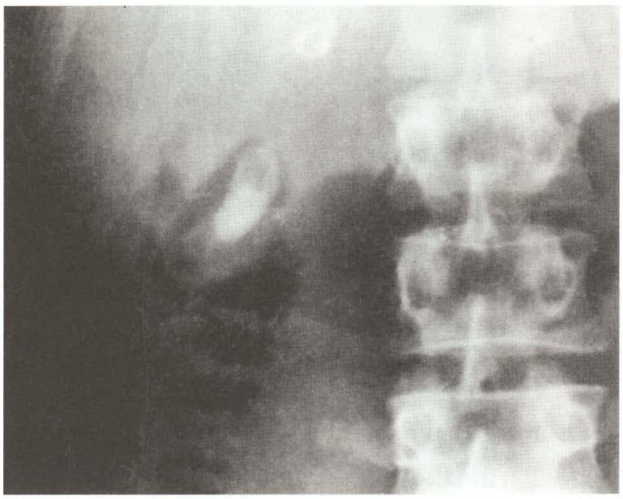


Fig. 50.4: Plain X-ray abdomen showing radio-opaque shadow in the region of gall bladder

4. What are the causes of radio-opaque shadow in the abdomen?

- Gall stones
- Renal stones
- Pancreatic stones
- Renal tuberculosis
- 'Chip' fracture of the transverse process of the vertebrae
- Calcified lymph nodes—tuberculosis
- Faecoliths
- Phleboliths

5. What is the treatment of symptomatic gall stones?

- Cholecystectomy

V. X-RAY CHEST PA VIEW SHOWING MULTIPLE ROUND SHADOW IN BOTH LUNG FIELDS

1. What is the diagnosis?

- Bilateral chest secondaries

2. What are they called?

- Cannon ball secondaries

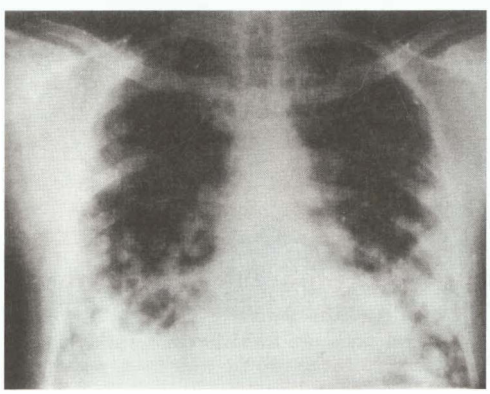


Fig. 50.5: X-ray chest PA view showing multiple round shadows in both lung fields

3. Why are secondaries in the lung round?

- Lung is an elastic tissue, it has resilience. Hence, durin the act of inspiration and expiration, equal amount o pressure is exerted on secondaries, which are growing Hence, they tend to become round.

4. What are the common causes of chest secondaries?

1. Carcinoma breast
2. Carcinoma testis
3. Malignant melanoma
4. Hepatoma
5. Renal cell carcinoma
6. Sarcoma

5. Is there any other differential diagnosis?

- Miliary tuberculosis: The shadows will be very small anc numerous.

VI. X-RAY CERVICAL VERTEBRAE WITH UPPER RIBS SHOWING BILATERAL CERVICAL RIBS

1. What is a cervical rib?

- It is an extra rib arising from 7th cervical vertebra.

2. What are 4 types of cervical rib?

- Incomplete bony
- Complete bony with anterior expanded bony end.
- Partly fibrous, partly bony
- Complete fibrous band

3. What variety gives rise to vascular symptoms?

- The fibrous band variety

4. What is your finding here?

- On the right side, it is complete variety and on the left side, it is incomplete.

5. If cervical rib is symptomatic, what is the treatment?

- Extraperiosteal excision of cervical rib which means removal of the rib along with the periosteum. Some surgeons also do cervical sympathectomy to decrease vasomotor tone of vessels.

Fig. 50.6: X-ray cervical vertebrae with upper ribs showing bilateral cervical ribs

VII. BARIUM SWALLOW SHOWING INTRINSIC, IRREGULAR, AND PERSISTENT FILLING DEFECT IN THE LOWER OESOPHAGUS

1. What is the diagnosis?

- Carcinoma lower one-third of oesophagus.

2. What are the other findings?

- Proximal shouldering is very characteristic of malignancy.

3. How do you confirm the diagnosis?

- Oesophagoscopy and biopsy

4. If biopsy report is adenocarcinoma, what is the treatment?

- Operable—oesophagogastrectomy
- Inoperable—to relieve dysphagia, metallic stents can be introduced. Thus, surgery can be avoided.

5. What are the premalignant conditions?

- Achalasia cardia
- Reflux oesophagitis
- Corrosive stricture
- Plummer-Vinson syndrome

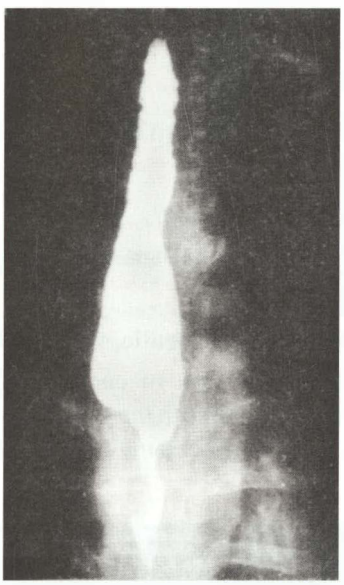


Fig. 50.7: Barium swallow showing intrinsic, irregular and persistent filling defect in the lower oesophagus

VIII. BARIUM SWALLOW SHOWING EXTENSIVE AND IRREGULAR FILLING DEFECT INVOLVING MIDDLE ONE-THIRD OF OESOPHAGUS

1. What is the diagnosis?

- Carcinoma middle one-third of oesophagus

2. How do you confirm the diagnosis?

- Oesophagoscopy and biopsy

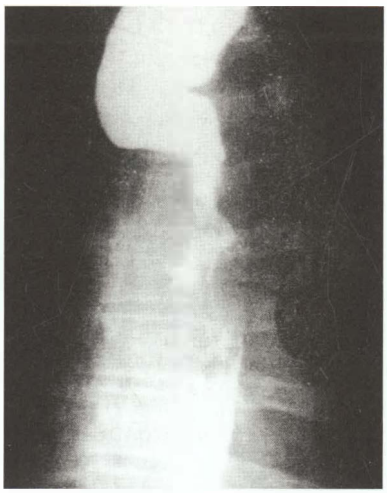


Fig. 50.8: Barium swallow showing extensive and irregular filling defect involving middle one-third of oesophagus

3. What will be the biopsy report?

- Squamous cell carcinoma

4. What other investigations are necessary in such case?

- Bronchoscopy, CT scan of the chest and endosonography are important investigations.

5. Looking at this advanced lesion, what is probably the best treatment for this patient?

- Radiotherapy followed by dilatation of the oesophagus, since chances of fibrosis and narrowing of the lumen following radiotherapy are high.

IX. BARIUM MEAL SHOWING INTRINSIC, IRREGULAR, AND PERSISTENT FILLING DEFECT INVOLVING PYLORIC ANTRUM

1. What is the diagnosis?

- Carcinoma pyloric antrum

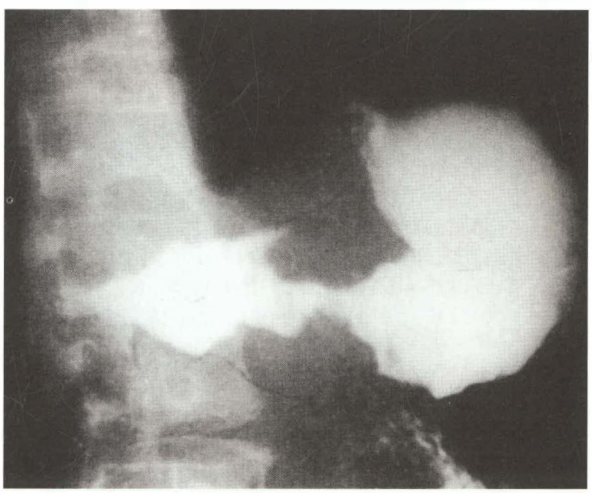


Fig. 50.9: Barium meal showing intrinsic, irregular and persistent filling defect involving pyloric antrum

2. How do you confirm diagnosis?

- Gastroscopic biopsy

3. What will be the biopsy report?

- Adenocarcinoma

4. What is the treatment, if it is operable?

- Subtotal gastrectomy

5. What structures are removed in the operation?

- Growth along with 60–70% of distal stomach, omentum, enlarged regional nodes such as prepyloric, suprapyloric, infrapyloric, left and right gastric nodes are removed, followed by gastrojejunal anastomosis.

X. BARIUM MEAL X-RAY SHOWING ENORMOUS DILATATION OF THE STOMACH AND FAILURE OF BARIUM TO FILL INTO THE DISTAL INTESTINE

1. What is the diagnosis?

- Gastric outlet obstruction due to chronic cicatrised duodenal ulcer (pyloric stenosis is an old terminology).

2. Why is it not due to carcinoma pyloric antrum?

- There is no filling defect in the pyloric antrum.

3. How do you treat this case?

- With a preoperative stomach wash, adequate intravenous fluids, total truncal vagotomy with GJ is the treatment of choice.

4. Why GJ and vagotomy?

- After vagotomy, motility of the stomach is lost and in pyloric stenosis, there is already obstruction at the pyloric antrum. Hence, gastrojejunostomy is the drainage procedure o choice.

5. Why not pyloroplasty or highly selective vagotomy?

- Pylorus is scarred and deformed. Hence, it is not safe to dc pyloroplasty. HSV is contraindicated in the presence o pyloric obstruction.

XI. BARIUM ENEMA SHOWING THE LEFT COLON, TRANSVERSE COLON AND A PART OF ASCENDING COLON

1. What is the diagnosis?

- Ileocolic intussusception

2. Why do you say so?

- The 'claw' like ending or pincer ending is typical of intussusception.

3. What are the causes of intussusception in adults?

- Submucous lipoma, or polyps
- Meckel's diverticulum
- Growth in the caecum
- Leiomyoma of the ileum

4. In a child, what are the causes?

- Weaning of the diet or viral infection.

5. What is the treatment of adult intussusception?

- Resection because there is a precipitating cause.

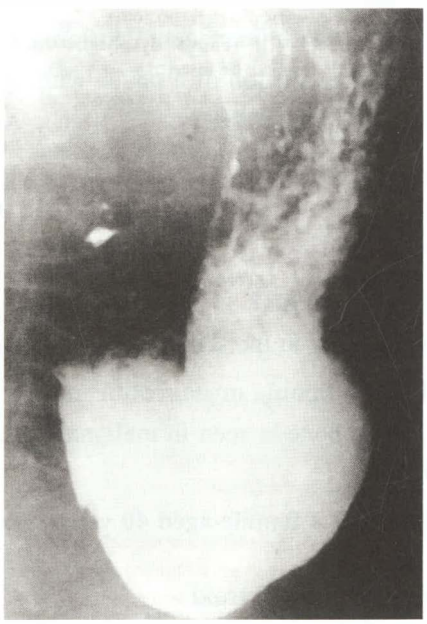


Fig. 50.10: Barium meal X-ray showing enormous dilatation of the stomach and failure of barium to fill into the distal intestine

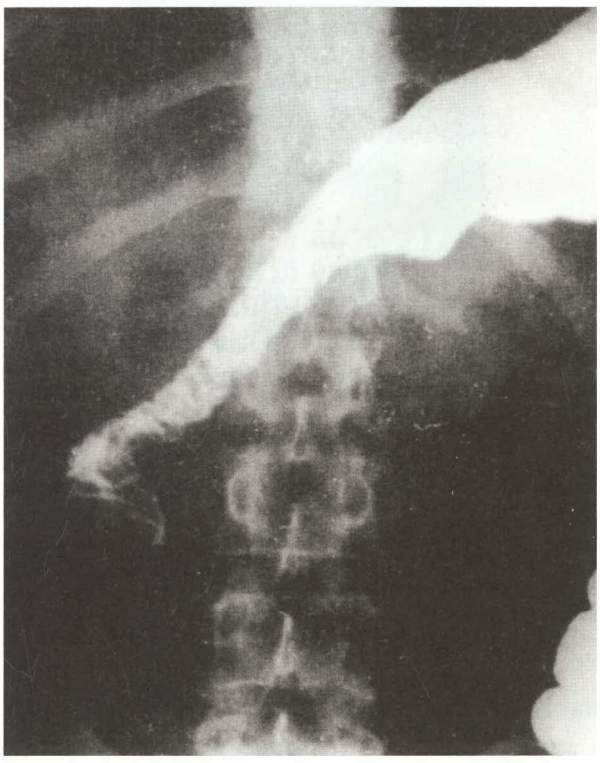


Fig. 50.11: Barium enema showing the left colon, transverse colon and a part of ascending colon with pincer ending

XII. BARIUM ENEMA SHOWING INTRINSIC, IRREGULAR AND PERSISTENT FILLING DEFECT IN THE ASCENDING COLON

1. What is the diagnosis?

- Carcinoma ascending colon

2. What is the confirmatory investigation?

- Colonoscopy and biopsy

3. What is the report, if it is carcinoma?

- Adenocarcinoma

4. What is the treatment?

- Right radical hemicolectomy, if it is operable. Structures removed in this operation include terminal ileum (6–8 cm), caecum including appendix, ascending colon and 1/3rd of right transverse colon. If it is inoperable, part of ileum is anastomosed to the transverse colon to prevent or relieve intestinal obstruction (side to side). One need not remove two feet of ileum.

5. What is the differential diagnosis?

- Ileocaecal tuberculosis: In this condition:
  1. Irregular filling defect is not seen.
  2. Caecum is usually pulled up and then ileocaecal angle becomes obtuse.

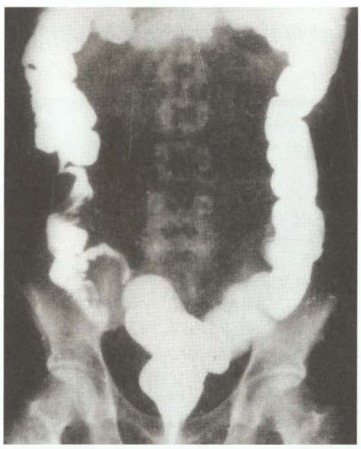


Fig. 50.12: Barium enema showing intrinsic, irregular and persistent filling defect in the ascending colon

XIII. BARIUM ENEMA SHOWING LOSS OF HAUSTRATIONS IN THE LEFT COLON, SMALL AND MULTIPLE, REGULAR FILLING DEFECTS DUE TO PSEUDOPOLYPOSIS

1. What is the diagnosis?

- Ulcerative colitis

2. What is pseudopolyposis?

- An attempt at healing in between the ulcers produces granulation tissue which have the appearance of polyps. Hence, pseudopolyposis.

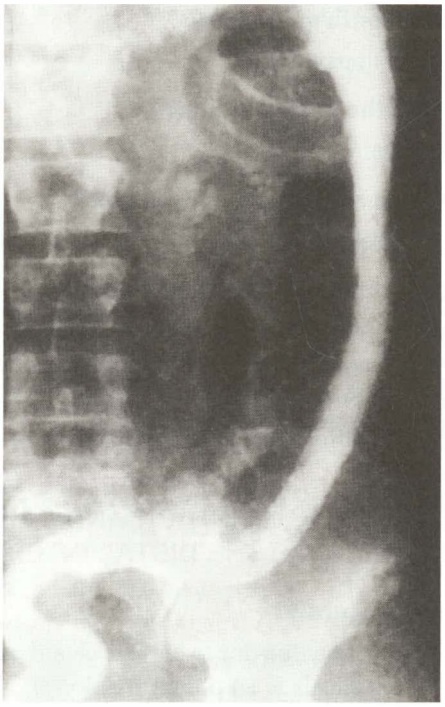


Fig. 50.13: Barium enema showing loss of haustrations in the left colon, small and multiple, and regular filling defects due to pseudopolyposis

3. What are the dangerous complications of ulcerative colitis?

- Haemorrhage, toxic megacolon, perforation and malignancy.

4. What are the drugs used in the treatment of ulcerative colitis?

- Salazopyrines and corticosteroids

5. What are the surgical treatments?

- Total colectomy with permanent ileostomy. OR
- Total colectomy, creation of a pouch with anastomosis of the pouch to the anal canal.

XIV. X-RAY LATERAL VIEW OF THE SKULL SHOWING A LARGE SWELLING WITH EROSION IN THE PERICRANIUM

1. What is the diagnosis?

- Secondary deposit in the skull

2. Why is it not a lipoma or neurofibroma?

- Erosion of the bone is seen in malignancy, not in benign tumours.

3. If this patient is a female aged 40 years, what are the causes?

- Follicular carcinoma thyroid
- Carcinoma of the breast
- Renal cell carcinoma

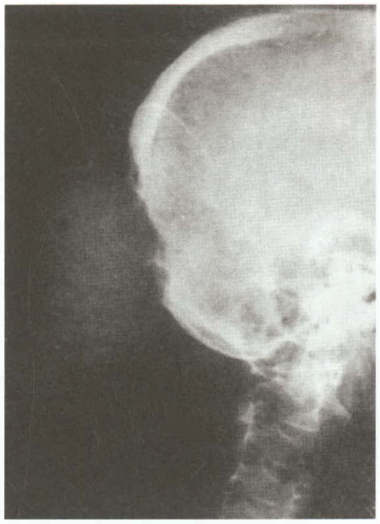


Fig. 50.14: X-ray lateral view of the skull showing a large swelling with erosion in the pericranium

4. What is the treatment if this is follicular carcinoma thyroid?

- Near total/total thyroidectomy followed by radio-iodine therapy and external radiotherapy for metastasis in bones.

5. How do you diagnose follicular carcinoma thyroid histologically?

- Angioinvasion and capsular invasion.

XV. ENDOSCOPIC RETROGRADE CHOLANGIO-PANCREATOGRAPHY (ERCP) SHOWING THE BILIARY AND PANCREATIC SYSTEMS

1. What is the diagnosis?

- Chronic pancreatitis

2. Why do you say so?

- Extensive calcification involving head, body and tail of pancreas.

Fig. 50.15: Endoscopic retrograde cholangiopancreatography (ERCP) showing the biliary and pancreatic systems

3. What is the simple investigation to diagnose chronic pancreatitis?

- Plain X-ray abdomen showing calcification

4. Why is ERCP done in this patient?

- To know whether the pancreatic duct is dilated or not.

5. If pancreatic duct is dilated more than 8 mm in a patient with severe abdominal pain with chronic pancreatitis, what is the treatment?

- Longitudinal pancreaticojejunostomy—Puestow's operation. In this operation, pancreatic duct is laid open, strictures are divided and the duct is anastomosed to jejunum.

XVI. T-TUBE CHOLANGIOGRAPHY SHOWING A FILLING DEFECT IN THE LOWER END OF THE COMMON BILE DUCT (CBD)

1. What is the diagnosis?

- Postcholecystectomy—residual stone in the CBD

2. What is the surgery done for this patient?

- Cholecystectomy and choledocholithotomy

3. Why do you insert a T-tube after CBD exploration?

- In case of distal obstruction by a residual stone, the bile starts leaking from the suture line on the CBD and may result in biliary peritonitis. In such situations, T-tube helps in drainage of the bile.

4. What material is T-tube made of?

- Latex

5. How do you treat this patient in order to extract the stone?

- Endoscopic sphincterotomy and extraction of the stone.

Fig. 50.16: T-tube cholangiography showing a filling defect in the lower end of the common bile duct (CBD)

XVII. SPLENOPORTOVENOGRAPHY (SPV) SHOWING EXTENSIVE COLLATERALS IN THE REGION OF SPLEEN

1. What is the diagnosis?

- Portal hypertension

2. What is the type of portal hypertension?

- Hepatic type

3. Why do you say so?

- Splenic vein, portal vein and its branches within the liver are visualised.

4. What is the probable cause in this patient?

- Cirrhosis of the liver

5. What is the first line of specific treatment for bleeding oesophageal varices?

- Endoscopic banding/sclerotherapy
- Banding is costly and needs more expertise.
- Sclerotherapy can be perivariceal or intravariceal-injection of 2% solution of sodium tetradecyl sulphate.
- Sclerotherapy is given at multiple sites and in multiple sittings.

Fig. 50.17: Splenoportovenography (SPV) showing extensive collaterals in the region of spleen

XVIII. RETROGRADE ANGIOGRAPHY SHOWING OCCLUSION OF FEMORAL ARTERY ON THE LEFT SIDE

1. What is the technique employed in this angiography?

- Seldinger's technique—percutaneous, transfemoral, retrograde.

2. What is the probable cause in our country?

- Buerger's disease (thromboangiitis obliterans).

3. Why do you say so?

- Buerger's disease affects medium-sized vessels and narrowing of femoral artery is segmental in this radiograph.

Fig. 50.18: Retrograde angiography showing occlusion of femoral artery on the left side

4. What is the surgical treatment for Buerger's disease?

- Lumbar sympathectomy

5. How does lumbar sympathectomy help these patients?

- By reducing the sympathetic tone of the lower limb, arterioles and capillaries get dilated allowing cutaneous ulcers to heal.

XIX. CONTRAST ENHANCED (CE) CT ABDOMEN SHOWING MASS IN THE RIGHT ILIAC FOSSA

1. What is this investigation?

- Contrast enhanced CT scan

2. How to interpret the CT scan?

- Structures imaged appear as densely white or black.

3. What is the name used to the picture in terms of number of units?

- Hounsfield units

4. Why do you give contrast?

- This is to increase the density between various structures. Example: Aorta appears bright with contrast.

5. What are the precautions?

- Pregnancy is a contraindication. Iodine containing contrast can give rise to nephropathy. Allergy to contrast can happen. Hence, dehydration should be corrected. Serum creatinine should be checked before contrast.

6. When do you use oral contrast?

- While studying abdominal viscera, e.g. if leak is suspected.

7. What is the finding here?

- It is showing a hypodense lesion in the right iliac fossa

8. What is the diagnosis?

- Mostly carcinoma caecum

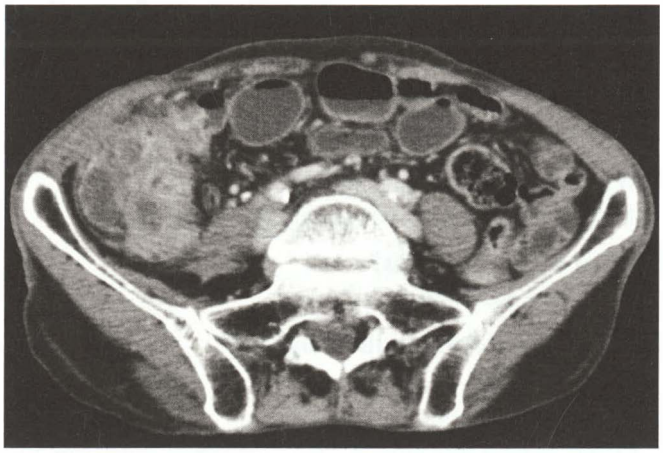


Fig. 50.19: A hypodense mass in the right iliac fossa involving caecum

9. Why do you say so?
- Anatomically it is a lesion occupying the right iliac fossa involving caecum.

10. How do you describe this?
- It is a hypodense mass with solid and cystic areas. Cystic areas represent tumour degeneration.

11. What else is seen in this picture?
- Fat planes between the mass and the abdominal wall is obliterated.

12. What is the importance of that?
- Probably it is infiltrating the abdominal wall.

13. Why do you want to know this information?
- At surgery, the involved portion of the abdominal wall has to be removed.

14. How do you confirm the diagnosis?
- Colonoscopy and biopsy

15. What will be the report expected?
- In majority of the cases it is adenocarcinoma

16. What is the treatment if it is operable?
- Right radical hemicolectomy

17. If it is inoperable, what is the treatment?
- Palliative ileo-transverse anastomosis

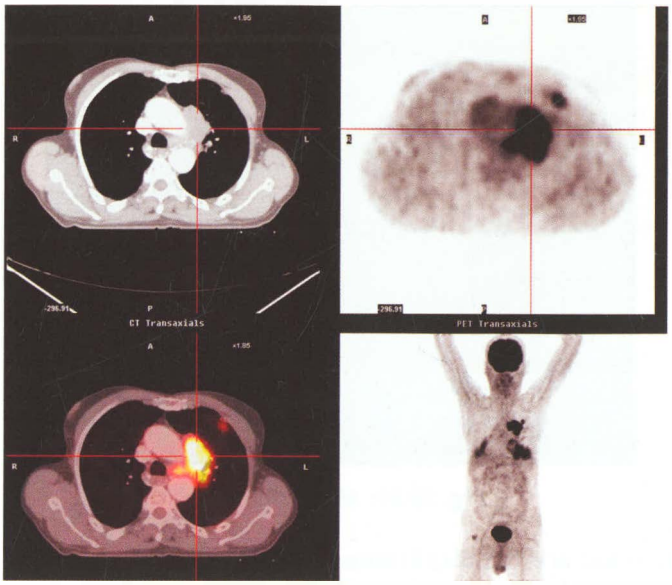
XX. POSITRON EMISSION TOMOGRAPHY (PET) SCAN

1. Name this investigation.
- PET-CT scan

2. What is PET scan?
- Positron emission tomography

3. What is the most commonly used positron emitting radionuclide?
- Fluoro-deoxyglucose (FDG)

4. What are the chief uses of PET scan?
- For myocardial perfusion and viability, detection of metastasis from cancer—carcinoma lung, colon, nasopharynx, etc.



PET-CT of a patient who was diagnosed to have carcinoma lung on bronchoscopy.
PET CT shows a hilar mass with a nodule anteriorly on left side of pleura.
It also shows pneumonic patch on lower zone of left lung which is FDG avid

Fig. 50.20: CT and PET of lung

5. What are the disadvantages?
- Very expensive and limited availability

6. What does this picture show?
- Hilar mass with a nodule anteriorly on the left side of pleura

7. What may be the diagnosis?
- Carcinoma lung

8. How do you confirm the diagnosis?
- Bronchoscopy and biopsy

9. If the report is adenocarcinoma lung, what is the next step?
- To stage the disease by whole body bone scan, PET scan, and CT scan.

10. If confined to lung, what is the treatment?
- Lobectomy/pneumonectomy.

XXI. MRI OF THE THIGH

1. Name this investigation
- Magnetic resonance imaging

2. What are the principles of MRI?
- Certain atomic nuclei, which possess unpaired protons or neutrons, possess an inherent spin. The nucleus is positively charged and therefore creates a small magnetic field around itself, when it spins. The human body contains in abundance such spinning nuclei in the atoms of hydrogen, which is found in water and lipids.

3. What are the chief advantages of MRI over CT scan?
- It is noninvasive and does not involve the use of ionising radiation. Hence, it is safe.

Fig. 50.21: MRI of the thigh

4. What are the disadvantages of MRI?
- The imaging time is long. Hence, movement of the patients may produce artefacts.
- Expensive
- Patients with pacemakers, metallic implant and critically ill patients cannot be scanned.
- Claustrophobia

5. What does this picture show?
- A hyperintense mass occupying the thigh region.

6. What is the diagnosis?
- Soft tissue sarcoma

7. How do you confirm the diagnosis?
- Trucut biopsy

8. Why not FNAC?
- FNAC cannot diagnose the type of sarcoma

9. What are common tumours in this location?
- Malignant fibrous histiocytoma (MFH) and liposarcoma.

10. What is the treatment?
- Wide excision with 2–3 cm margin

Please note: X-rays discussed here are the common X-rays which are asked in the MBBS examination. However, modern investigations such as mammogram, ultrasound, CT scan and MRI also may be asked. You are requested to read the chapters on radiology and breast for more details about these investigations.

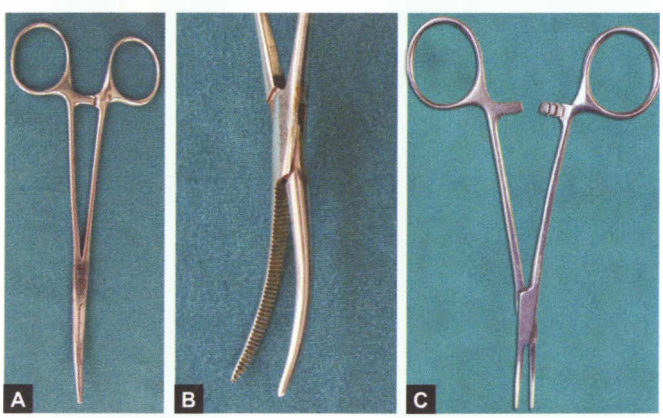
51

Instruments

- Forceps
- Retractors
- Occlusion clamps
- Dilators
- Tracheostomy tube
- Rubber tubes
- Catheters
- Sengstaken tube
- What is new?/Recent advances

ARTERY FORCEPS (HAEMOSTAT)

- It is also called **Spencer Well's artery forceps**. It has a ratchet and two blades with uniform serrations.
- It is used to control bleeding, not only from arteries but also from veins and capillaries. Once the bleeding points are caught, they are coagulated or ligature is applied.
- The curved artery is commonly used (Fig. 51.1B).
- The smaller version of this is called mosquito forceps (Fig. 51.1A). This is extremely useful in repair of harelip, cleft palate or other plastic surgery operations.
- It is also available as straight artery which is used to hold the stay sutures (Fig. 51.1C).



Figs 51.1A to C: (A) Mosquito forceps, (B) Curved artery forceps, (C) Straight artery forceps

ALLIS TISSUE HOLDING FORCEPS (Fig. 51.2)

- It has a ratchet and triangular expansion at the tip, where serrations are present.
- It can be used to **hold tough structures** such as fascia, aponeurosis, etc.
- Even though it can cause trauma, because of its better grip, it can be used to hold the duodenum for duodenal closure during gastrectomy.

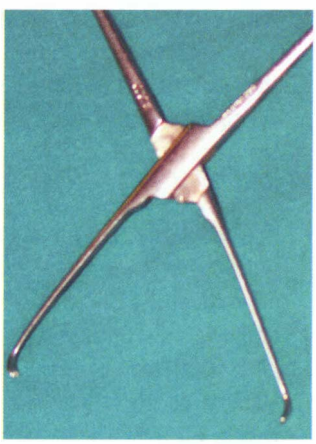


Fig. 51.2: Allis forceps

KOCHER'S FORCEPS (Fig. 51.3 and Key Box 51.1)

- This is similar to an artery forceps with serrations. It is available as curved and straight.
- There is a sharp tooth at the tip of the instrument. Hence, it has a better grip.
- Kocher's forceps can be used to **hold tough structures** like **aponeurosis, fascia**, etc.
- During thyroidectomy, it can be used to hold the strap muscles for dividing them.
- Theodor Kocher, a German surgeon, got the Nobel prize for his contribution to thyroid surgery.

KEY BOX 51.1

REMEMBER

- Kocher's forceps
- Kocher's test
- Kocher's thyroid dissector
- Kocher's vein
- Kocher's subcostal incision
- Kocher's gland holding forceps

SINUS FORCEPS (Fig. 51.4)

- This is like an artery forceps which has **No ratchet**.
- Serrations are confined to the tip so as to hold the wall of an abscess cavity, for biopsy.
- In ***Hilton's method*** of drainage of an abscess, once the incision is made, the sinus forceps is thrust into the abscess cavity and by opening the blades in all directions, the loculi are broken. To facilitate free opening of the blades, sinus forceps has no ratchet.

SWAB HOLDING FORCEPS (Fig. 51.5)

- This has a ratchet and two long blades
- Operating end is rounded with serrations
- It is used to hold the swab (gauze pieces) to prepare the parts with antiseptic agents at the time of surgery.
- This instrument can also be used as a blunt 'dissector' with the swab, while dissecting at a depth, e.g. lumbar sympathectomy, vagotomy.

BABCOCK'S FORCEPS (Fig. 51.6)

- An instrument with a ratchet and a triangular expansion with fenestrations at the operating end. It does not have any teeth. Thus, it is used to hold intestines during anastomosis or resection.
- This instrument can also be used to hold many other structures such as thyroid gland, mesoappendix, uterine tubes, etc.

LANE'S FORCEPS (Fig. 51.7)

- This is similar to Babcock's forceps but the tip is more broad, expanded with a bigger opening.
- It is used to hold the appendix
- However, it does not seem to have any additional advantage when compared to Babcock's forceps.

DISSECTING SCISSORS

- This is also called **Mayo's scissors** (Fig. 51.8 and Key Box 51.2).
- It does not have ratchet and operating end is sharp
- This is used to dissect tissue planes during surgical operations and to cut or divide important structures.
- It is popularly called **tissue scissors**.

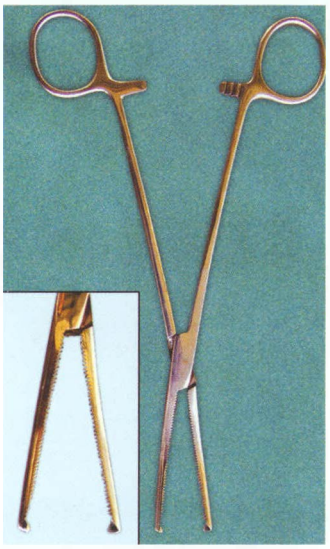


Fig. 51.3: Kocher's forceps

Fig. 51.4: Sinus forceps

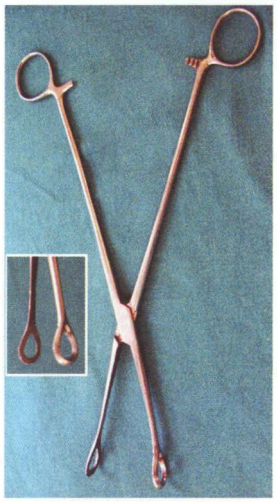


Fig. 51.5: Swab holding forceps

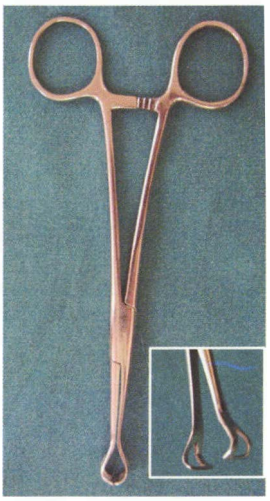


Fig. 51.6: Babcock's forceps

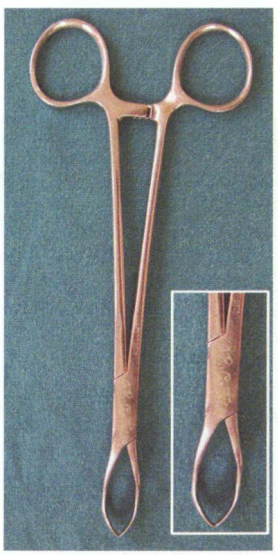


Fig. 51.7: Lane's forceps

Fig. 51.8: Mayo's scissors

KEY BOX 51.2

REMEMBER

- Mayo's scissors
- Mayo's herniorrhaphy
- Mayo's posterior GJ
- Mayo's vein
- Mayo's needle (used for hernia repair)

STRAIGHT SCISSORS (Fig. 51.9)

It is used to cut the sutures or knots. Hence, called suture-cutting scissors.

DISSECTING FORCEPS (Fig. 51.10)

- This is a toothed forceps. It is also available as non-toothed forceps.
- Dissecting forceps with dissecting scissors makes good 'tool' for a surgeon to develop a tissue plane in majority of surgeries.
- The forceps is very useful to 'pick' individual layers such as serosa, seromuscular layers, mucosa, etc. during anastomosis.

NEEDLE HOLDER (Fig. 51.11)

- This is a long instrument with a ratchet at non-operating end.
- The operating end has two small blades with serrations.
- The instrument is used to **hold the curved needles** which are used to suture the parts.
- A firm grip is essential to apply proper sutures.

SCALPEL WITH BLADE (Fig. 51.12)

- This is popularly called **surgeon's knife.**
- This is used to incise the skin and subcutaneous tissue.
- Due to the sharp nature, it can be used to divide a major vascular pedicle once ligatures are applied.

CHEATLES FORCEPS (Fig. 51.13)

- It is a long instrument having a curved shaft
- The **handle has no lock**
- It is kept dipped in antiseptic solutions
- This instrument is used to pick up sterilised articles such as sponges, gauze pieces or other instruments and to transfer to the instrument trolley.

DEAVER RETRACTOR (Fig. 51.14)

- This is popularly called **Deaver liver retractor**
- It has a long blade and operating end is curved
- It can be used to retract the liver during vagotomy, cholecystectomy or gastrectomy, etc.

Fig. 51.9: Straight scissors

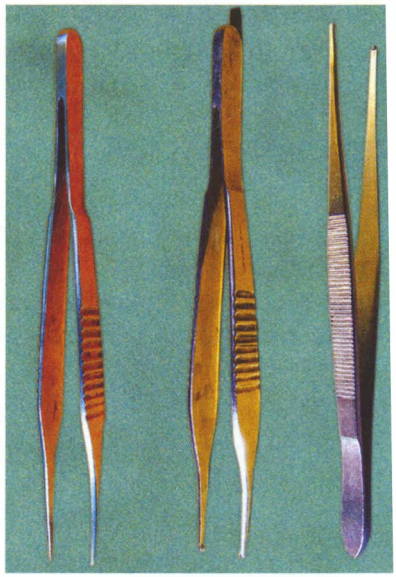


Fig. 51.10: Dissecting forceps

Fig. 51.11: Needle holder

Fig. 51.12: Scalpel with blade

Fig. 51.13: Cheatles forceps

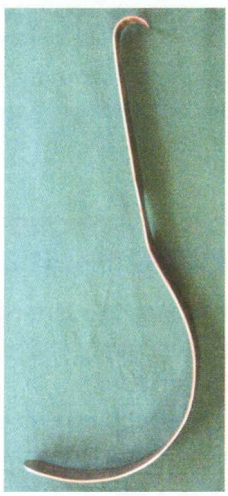


Fig. 51.14: Deaver retractor

- Since it has long blades, it can be used to retract the kidney upwards, during lumbar sympathectomy or to retract the urinary bladder during surgery on the rectum.

MORRIS RETRACTOR (Fig. 51.15)

- This is a long instrument with broad operating end.
- This is used **to retract the abdominal wall**, once the peritoneum is opened.
- However, if a self-retaining retractor is used to widen the laparotomy wound, the use of Morris retractor gets limited.

Fig. 51.15: Morris retractor

CZERNY RETRACTOR (Fig. 51.16)

- This is a double-hooked retractor on one side and a single blade on the other side.
- This is a **superficial retractor**, can be used to retract layers of the abdominal wall, muscles, etc. Thus, during appendicectomy, herniorrhaphy or thyroidectomy, this instrument is very useful.

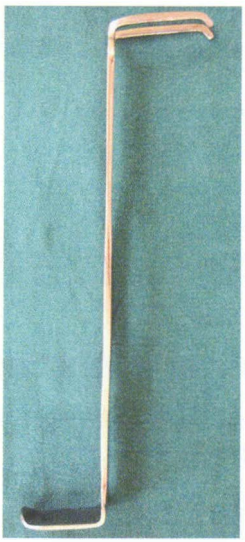


Fig. 51.16: Czerny retractor

LANGENBECK RETRACTOR (Fig. 51.17)

- This instrument has only **one blade**
- The uses of this are similar to that of Czerny's retractor.

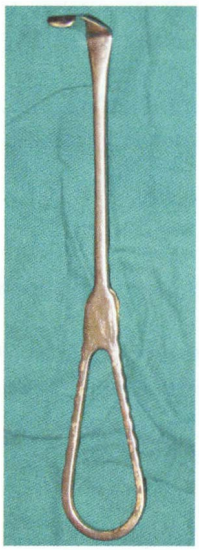


Fig. 51.17: Langenbeck retractor

MOYNIHAN'S STRAIGHT OCCLUSION CLAMP (Fig. 51.18)

- This is a long instrument with a ratchet. The operating end has two long blades with serrations in the line of blades.
- This instrument is used to **occlude the intestinal lumen** to prevent spillage of intestinal contents during intestinal resection or intestinal anastomosis.
- This does not interfere with the vascularity of the intestine.

Fig. 51.18: Moynihan's occlusion clamp

PAYR'S CRUSHING CLAMP (Fig. 51.19)

- This is a heavy instrument with **double lever system**, because of which it has a better grip
- The two short blades have uniform serrations
- During gastrectomy, when portion of the stomach is excised, this instrument is applied on the stomach side so that the stomach with this instrument is excised.

Fig. 51.19: Payr's crushing clamp

DESJARDIN'S CHOLEDOCHOLITHOTOMY FORCEPS (Fig. 51.20)

- This is a long curved instrument with no **ratchet**
- The operating end is expanded with fenestrations
- The tip is blunt
- It is used to **extract stones from common bile duct**. It can also be used to extract stones from the ureter.
- Since there is no ratchet, free opening is possible, and the stones do not get crushed.

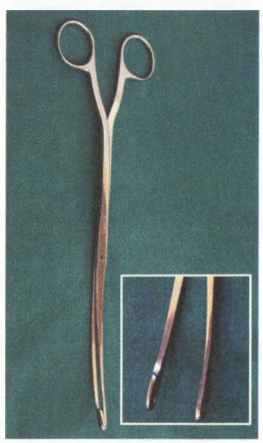


Fig. 51.20: Desjardin's forceps

BAKE'S DILATOR (Fig. 51.21)

- This is a long malleable instrument available in various diameters.
- It has a handle, long body and the tip is blunt
- Once common bile duct exploration is completed, this dilator is passed, to assess for any distal obstruction.
- The free passage of Bake's dilators of different sizes indicate that there is no distal obstruction (however, to be confirmed by cholangiogram).

KOCHER'S THYROID DISSECTOR (Fig. 51.22)

- This has a long handle and the operating end is small and blunt with an opening.
- A few longitudinal serrations are present at the tip.
- This was used to dissect the upper pole of thyroid gland.
- This instrument can also be used to dissect the isthmus of the thyroid gland from the trachea.
- **Silk thread can be fed** into the opening so as to ligate the vascular pedicle or isthmus.
- With the availability of the right-angled forceps, this instrument is not in routine use nowadays.

ANEURYSM NEEDLE (Fig. 51.23)

- It is a long instrument with an ***eye*** at the operating end.
- It is called aneurysm needle because it was used to ligate the feeding artery in an aneurysm. However today, this instrument is of limited use.
- During **venesection or cut down**, the silk suture can be threaded within the ***eye***, passed round the vein and it is tied.

TROCAR AND CANNULA (Fig. 51.24)

- This has two parts. The inner sharp part is the trocar and outer blunt part is cannula.
- It is used to drain hydrocoele fluid.
- Once hydrocoele sac is delivered, it is punctured wit trocar and cannula, the trocar removed and the flui drained.
- Make sure that trocar and cannula should match, otherwis injury to the deeper structures (testis) can occur.

HUMBY'S KNIFE (Fig. 51.25)

- This instrument has a handle and a long sheath.
- When in use, a disposable blade can be attached to it.
- The instrument is used to take skin graft. Hence, it is alsc called skin grafting knife.
- To facilitate the exact thickness of the skin to be removed there is a screw at the operating end, with which, prio adjustment should be done.

MYER'S METAL STRIPPER (Fig. 51.26)

- This is a long metallic chain or a stripper used in varicose vein surgery.
- It has a handle which is T-shaped and the 'advancing' enc which enters the vein. This is blunt. Once this end comes out of the cut end of the vein, a medium-sized head is connected to it.
- With gentle force (traction) exerted on the handle, the varicose vein can be stripped.
- Hence, it is also called vein stripper

SELF-RETAINING RETRACTOR (Fig. 51.27)

- It is a strong, heavy instrument, with two blades.
- This is used to spread the laparotomy wound. Hence, it is called self-retaining retractor.

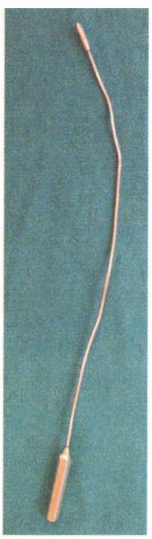


Fig. 51.21: Bake's dilator

Fig. 51.22: Kocher's thyroid dissector

Fig. 51.23: Aneurysm needle

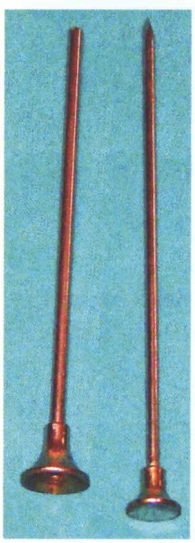


Fig. 51.24: Trocar and cannula

Fig. 51.25: Humby's knife

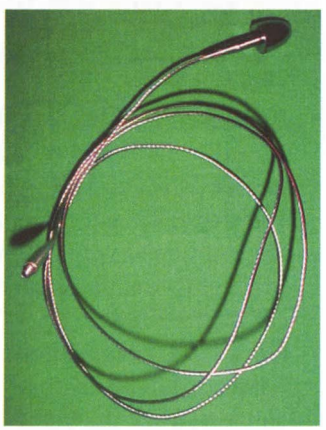


Fig. 51.26: Myer's metal stripper

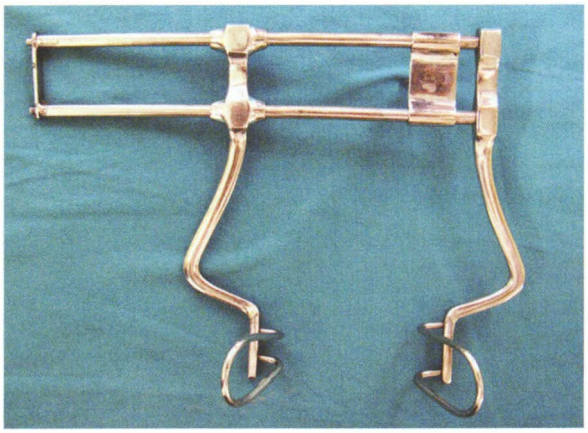


Fig. 51.27: Self-retaining retractor

RIB SPREADER (Fig. 51.28)

- This is also a strong heavy instrument with two long blades.
- Once an incision is deepened through the intercostal spaces and the pleura is opened, the rib spreader is used and by rotating the latch handle, the ribs are spread apart.

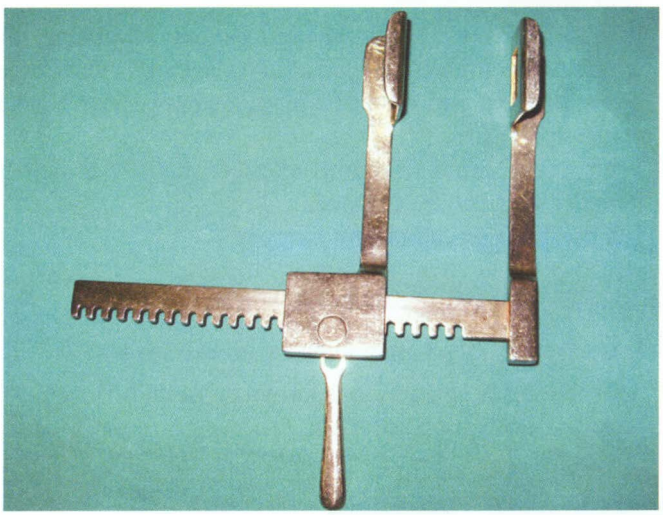


Fig. 51.28: Rib spreader

PROCTOSCOPE (Fig. 51.29)

- This is an instrument used to visualise the rectum and the anal canal.
- It has an outer sheath with the handle (A).
- An inner blunt part is called obturator (B).
- Before introducing the proctoscope one must make sure that obturator and the outer sheath must match. Lubricate the instrument well before introducing.
- In painful conditions such as fissure *in ano*, proctoscopy is contraindicated.
- Once rectal examination is done, proctoscope is held firmly with the left hand (buttocks separated), the obturator is supported by the right hand. The instrument is slowly introduced inside. The obturator is removed and rectum is visualised using light source.

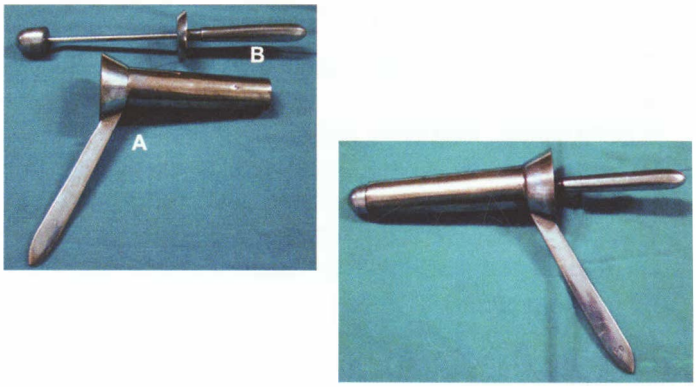


Fig. 51.29: Proctoscope

- Proctoscope is used to diagnose haemorrhoids, carcinoma rectum or rectal ulcers, etc. Biopsy can be taken with a biopsy forceps in nonhealing ulcers of the rectum. Haemorrhoids can be injected and pelvic abscess can be drained into the rectum with the help of a proctoscope.

LISTER'S METAL DILATOR (LISTER'S BOUGIE)

- This is a long instrument curved at the tip. Its diameter is written near the handle. It is available in various diameters. The difference between the two numbers is 3. The maximum size of the Lister's dilator is 9/12 (Fig. 51.30).
- The tip is olive-pointed and the end of the handle is round. The minimum and maximum diameter of the instrument is written on the handle. The other type of bougie is Glutton's bougie with a plain tip and the end of the handle is trapezoid. The maximum size of Glutton's bougie is 24/28 and difference between the two numbers is 4.

MALE METALLIC CATHETER (Fig. 51.31A)

- These catheters are used to drain urine in cases of retention of urine when rubber catheter fails.
- It is a long instrument which is curved because the male urethra is long and curved.
- It has two eyes at the distal end which are situated laterally and at different levels so that the instrument does not become weak at that spot.
- Once the urine is drained, the catheter can be left in place by passing a thread through the two rings present at the proximal end and fixing them to patient's thigh.
- Due to the fear of false passage, injury to the urethra and introducing infection, this catheter is not used nowadays. It is replaced by trocar suprapubic cystostomy.

FEMALE METALLIC CATHETER (Fig. 51.31B)

- Used to drain urine in females
- This is a short and straight instrument because the urethra is short and straight in females.

Fig. 51.30: Lister's dilator

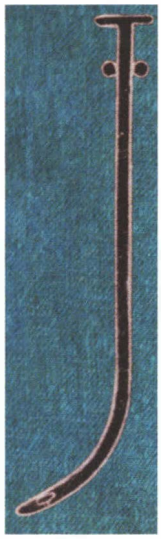


Fig. 51.31A: Male metallic catheter

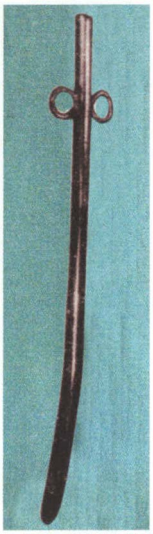


Fig. 51.31B: Female metallic catheter

Fig. 51.32: Towel clip

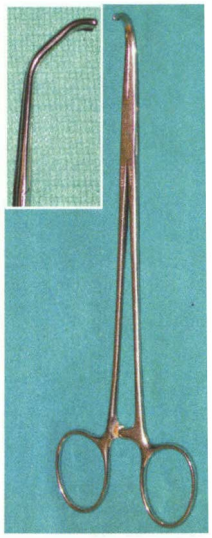


Fig. 51.33: Right-angled forceps

- It has multiple holes at the tip
- Indications for usage of this catheter are very rare because acute retention of urine is rare in females and even if it occurs, a red rubber catheter can be passed.
- It is used to empty bladder before vaginal hysterectomy and other gynaecologic surgeries.
- Emptying the bladder is mandatory before any gynaecological examination of a patient.

TOWEL CLIP (Fig. 51.32)

- This instrument has a ratchet and the operating end is sharp.
- This is available in different sizes
- Once the part is cleaned and draped the clips are used to hold the towels in place.

RIGHT-ANGLED FORCEPS: LAHEY'S FORCEPS

- This is a long instrument with right angle at the operating end (Fig. 51.33).
- This instrument is extremely useful in **ligating the major vascular pedicles**, e.g. superior thyroid pedicle—thyroidectomy.
- Cystic artery: Cholecystectomy
- Lumbar veins: Lumbar sympathectomy

HUDSON'S BRACE AND THE BURR (Fig. 51.34)

- This is a heavy instrument with a brace and the burr (drill).
- This is used to create **openings into the cranium** so as to get an access to the structures within.
- Thus once a 'burr' is made, drainage of blood, fluid or pus can be done.

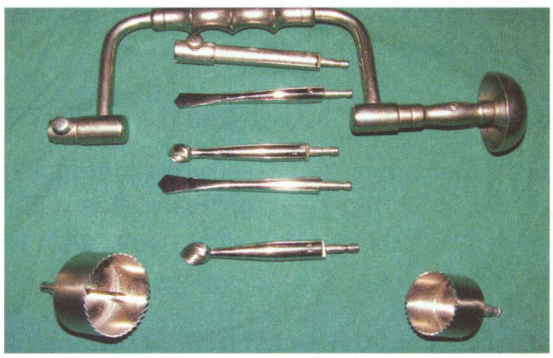


Fig. 51.34: Hudson's brace and the burr

CRICOID HOOK (Fig. 51.35)

- This has a broad handle and a thin shaft with a hook at the operating end.
- This is used to stabilise the trachea by hooking the cricoid cartilage 'up'.
- This step is essential in children wherein veins are very superficial and can get injured easily when child moves the head and neck. By stabilising the trachea, it is easy to incise the trachea, without injuring the vessels.

TRACHEAL DILATOR (Fig. 51.36)

- This is an instrument with **no ratchet at** the nonoperating end.
- The operating end is blunt and curved
- The peculiarity of this instrument is that **when the handle is opened, operating end is closed** and **when the handle is closed, operating end is opened**.
- Tracheal dilator is used in the **post-tracheostomy period**, when the tube has to be changed due to blockage. In such situations, once the tube is removed, tracheal dilator is

Fig. 51.35: Cricoid hook

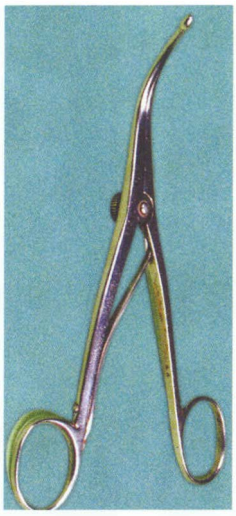


Fig. 51.36: Tracheal dilator

Fig. 51.37: Fergusson's amputation saw

Fig. 51.38: Bone nibbler

introduced, the opening in trachea is kept open, and the new tube is introduced. However, once the track is formed, tracheal dilator need not be used.

FERGUSSON'S AMPUTATION SAW (Fig. 51.37)

Amputation saw has teeth on its cutting edge to facilitate cutting through the bone and is of different sizes. They are manufactured with one- or two-sided cutting edge for limb amputations.

Uses: In lower limb, amputations commonly—above knee (AK) amputation and below knee (BK) amputation.

BONE NIBBLER (Fig. 51.38)

It is also called double action bone nibbler, identified by long handle and small jaws, top jaw is used for cutting and lower jaw is used to hold the tissue firmly.

Uses: To make cut end of the bone smooth after amputation rib cutting and to enlarge burr hole.

BONE FILE/RASPATORY (Fig. 51.39)

One side of the raspatory is used to hold as a handle, while its other side has sharp projections with both fine and coarse teeth on both sides with a flat blade.

Uses: Blunt separation of the periosteum and connective tissue from the surface of the bone, smoothening of sharp bony edges after amputation and before fixing fractures.

VOLKMANN CURETTE (Fig. 51.40)

The edges of the distal spoon-shaped part of this instrument are sharp which make it possible to remove the tissues.

Uses: Scoop the granulation tissue, to clean the base of the infected wound, and to remove the infected bone in the case of osteomyelitis.

DEBAKEY FORCEPS (BAYONET STYLE) (Fig. 51.41)

They are typically large—some examples are upwards of 12 inches (36 cm) long, and have a distinct coarsely ribbed grip panel, as opposed to the finer ribbing on most other tissue forceps, a type of atraumatic tissue forceps.

Uses: In vascular procedures to avoid tissue damage during manipulation. Less traumatic manipulation of tissue and used during suturing.

Fig. 51.39: Bone file

Fig. 51.40: Curette

Fig. 51.41: Debakey forceps

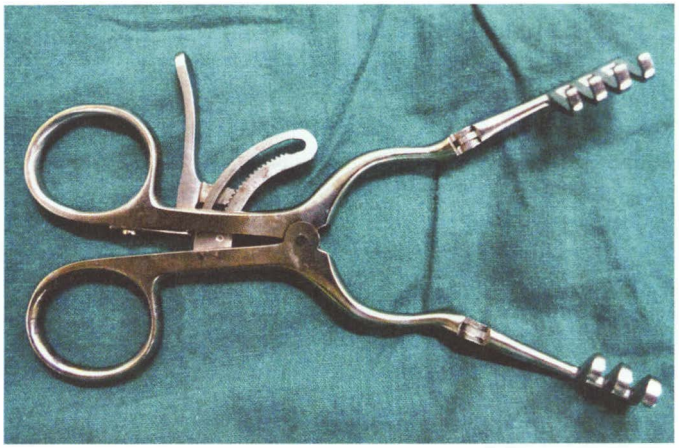


Fig. 51.42: Beckman-Adson laminectomy retractor

BECKMAN-ADSON LAMINECTOMY RETRACTOR (Fig. 51.42)

It has hinged blades with 4 × 4 prongs, an adjustable swivel arms and a ratchet to hold tissue apart.

Uses: Retraction in procedures involving deep tissues like in laminectomy for spinal surgeries.

RIBBON MALLEABLE RETRACTOR (Fig. 51.43)

A malleable or ribbon retractor (manual) may be bent to various shapes.

Uses: It is used at the end of the case to keep the viscera away during the fascial closure and is also used to retract deep wounds.

THE HARMONIC SCALPEL (Fig. 51.44)

The harmonic scalpel is a new device that has been introduced to surgery during the last decade. It is a device that uses high-frequency mechanical energy to cut and coagulate tissues at the same time.

It uses ultrasound technology to cut tissues while simultaneously sealing the edges of the cut.

Active tips of the harmonic scalpel employ a rigid active lower blade through which the vibrating energy is transmitted. The movable upper jaw is used to compress the vessel against the lower blade, thus allowing transfer of the vibrational energy.

The instrument is similar to an electrosurgery instrument and can be used in all open and laparoscopic surgeries, but superior in that it can cut through thicker tissue, creates less toxic surgical smoke and may offer greater precision especially during a laparoscopic surgery.

BIPOLAR CAUTERY (Fig. 51.45)

When the electric current is passing between the two parts of the instrument, we call it the bipolar diathermy/cautery (e.g. bipolar forceps).

Fig. 51.43: Malleable retractor

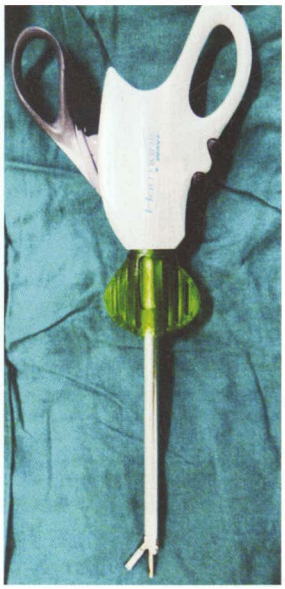


Fig. 51.44: Harmonic scalpel

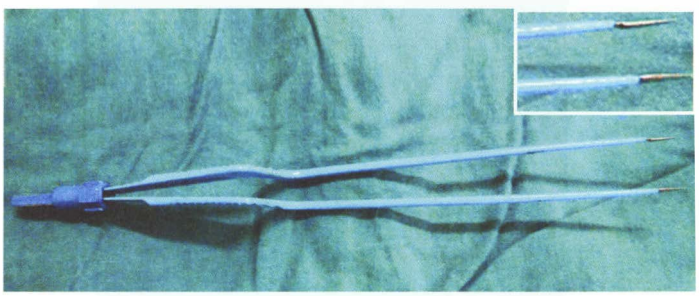


Fig. 51.45: Bipolar cautery

It makes possible to perform a more precise work and the size of the burned area is small and is more useful when haemostasis is required close to the nerves.

Uses: Thyroid surgery when close to RLN (recurrent laryngeal nerve) neurosurgery or spinal surgeries.

ALLISON'S LUNG RETRACTOR (Fig. 51.46)

It is retractor with a special type of blade, made of wires, in the form of a net over one end and a handle at the other end.

Uses: For retraction of the lung in thoracotomy. It does not damage the lungs and the lungs can expand in between the wires.

GIGLI SAW (Fig. 51.47)

Composed of a wire as a blade and two handles to hold the wire on either side.

Uses: For bone cutting in amputation surgeries similar to amputation saw such as below knee and above knee amputations commonly.

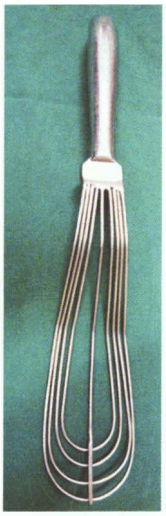


Fig. 51.46: Allison's lung retractor

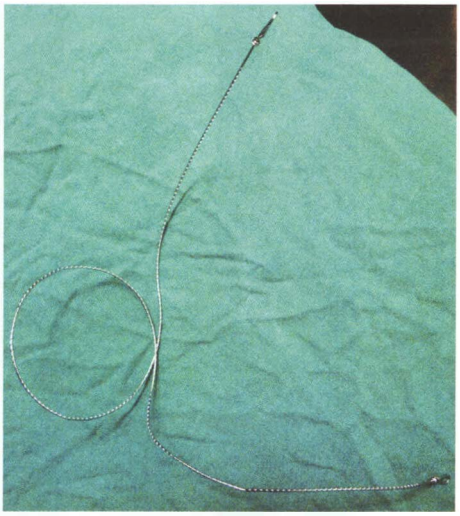


Fig. 51.47: Gigli saw

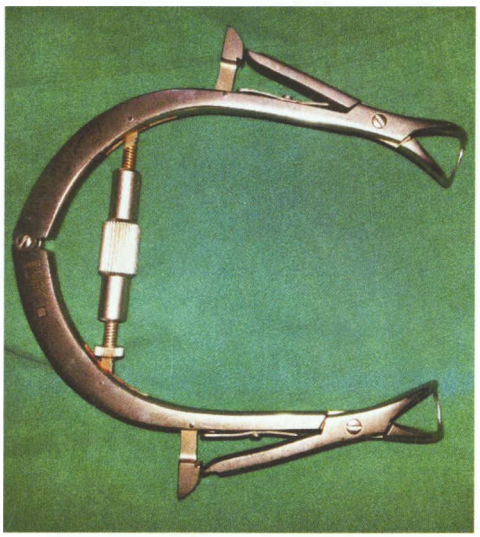


Fig. 51.48: Joll's retractor

JOLL'S THYROID RETRACTOR (Fig. 51.48)

It is a self-retaining retractor, which is held by the two towel clip like forceps on both sides to hold the flaps and can be adjusted using a screw in between.

Uses: To retract skin flaps during thyroid surgery.

METAL TRACHEOSTOMY TUBE (Fig. 51.49)

- This has **two tubes**, the inner long and the outer short tube.
- This has no **cuff**.
- Once the tube is introduced, the tape is passed around the neck, passed through the opening and tied so as to keep the tube in place.
- If the tube is blocked, the inner tube can be removed, cleaned and reintroduced.

Fig. 51.49: Metal tracheostomy tube

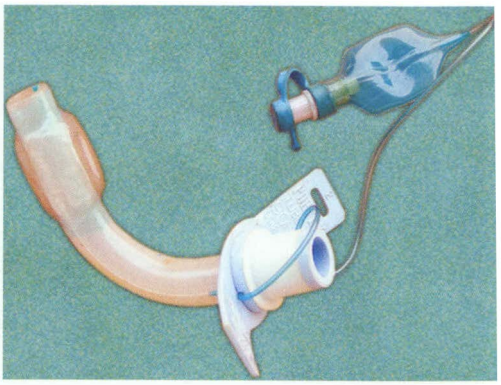


Fig. 51.50: Cuffed tracheostomy

- Metal tracheostomy tubes are useful as permanent tracheostomy tube.

CUFFED TRACHEOSTOMY TUBE (Fig. 51.50)

- This is made of polyvinyl chloride. It is a **single tube**.
- Once the tube is introduced within the trachea, the cuff is inflated by using 3–5 ml of air.
- The cuff prevents leakage of air and prevents **acid aspiration syndrome (Mendelson's syndrome)**.
- If this tube is blocked, it is an emergency. In such cases, the tube has to be cleaned and mucus plugs have to be removed. Otherwise, the tube is removed, the tracheal opening is kept open with the help of tracheal dilator and a new tube is introduced. Alternatively, endotracheal intubation may need to be done to ensure patency of the airway.

CORRUGATED RED RUBBER DRAIN (Fig. 51.51)

- It is made of red rubber. It has corrugations on both sides. Whenever a major surgery is done, some amount of blood loss or anastomotic leakage is expected. This drain is used so that **fluid can escape freely outside**.
- Thus, it is used after thyroidectomy, gastrectomy, cholecystectomy, etc. The drain is removed after it stops draining. Usually it takes about 3–5 days.
- After laparotomy for peritonitis, these drains are used to prevent residual abscess in the postoperative period.

MALECOT'S CATHETER (Fig. 51.52)

This is made of red rubber. It has flower-shaped end and has a wide diameter. It is used to drain amoebic liver abscess. It is straightened with the help of an introducer and left in cavity and brought outside. It is a self-retaining catheter. This is used to drain urinary bladder after transvesical prostatectomy or can be used as feeding gastrostomy tube. It can also be used to drain empyema thoracis.

Fig. 51.51: Corrugated red rubber drain

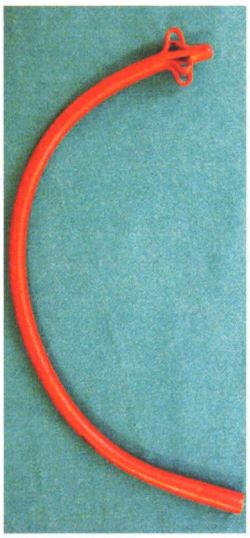


Fig. 51.52: Malecot's catheter

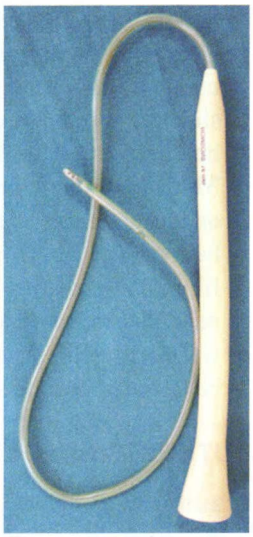


Fig. 51.53: Mousseau Barbin's tube

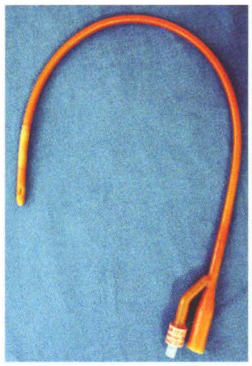


Fig. 51.54A: Foley catheter

Fig. 51.54B: Distended bulb

Fig. 51.55: Red rubber catheter

MOUSSEAU BARBIN'S TUBE (Fig. 51.53)

- This is also called MB tube. It is a funnel-shaped tube with Ryle's tube like attachment. It is used in inoperable cases of carcinoma oesophagus to palliate dysphagia. It is stitched to the Ryle's tube which is brought out through the mouth and it is slowly drawn in by pulling the other end of Ryle's tube which is in the stomach, after doing a gastrostomy.
- Once the tube is below the level of growth, it is cut at a sufficient distance and is stitched to the stomach wall.
- With the availability of laser coagulation of the growth, and considering discomfort caused by the tube including its migration, the MB tube is not popular and not preferred.

FOLEY'S SELF-RETAINING URINARY CATHETER (Fig. 51.54)

- This is made of **latex with silicon coating**. At the tip, there is a bulb, capacity of which is written at the other end.
- Before inflating the bulb, one must make sure that **catheter is in the urinary bladder, not in the urethra**. This is assessed by free flow of urine.
- After introducing the catheter, the bulb is inflated using saline. Thus, it becomes self-retaining. After the usage, it is removed by deflating the bulb. It can also be used to drain peritoneal cavity as in biliary peritonitis. Inflated bulb compresses the prostatic bed and controls bleeding after prostatectomy.

RED RUBBER CATHETER (Fig. 51.55)

This is used to drain urine temporarily. It causes urethritis if it is left long in the urinary bladder. Once the urine is emptied, it is removed. It is not a self-retaining catheter. Not routinely used nowadays because of availability of Foley's catheter. It is more stiff than Foley catheter. Hence, **in cases of stricture urethra where Foley's catheter cannot be passed, red rubber catheter may be used.**

NASOGASTRIC TUBE/ RYLE'S TUBE (Fig. 51.56)

Fig. 51.56: Ryle's tube

- This is also called **Naso-gastric tube**. At the end of this tube there are **lead shots.** After introducing within the stomach, its position is confirmed by pushing 5–10 ml of air and auscultating in the epi-gastrium or aspirating gastric juice. It is a long tube having 3 marks. When the tube is passed up to the 1st mark, it enters the stomach. Usually it is passed up to 2nd mark. Life-saving use of **Ryle's tube** is in **acute gastric dilatation.**
- **In volvulus of the stomach, it is impossible to pass a Ryle's tube.**
- Ryle's tube is used to decompress the stomach as in intestinal obstruction or pyloric stenosis.
- It is used in the diagnosis of GI haemorrhage
- It is also used to provide enteral nutrition to comatose patients or critically ill patients.

T-TUBE (KEHR'S) (Fig. 51.57)

- This is a flexible tube made of latex with a long vertical limb and a short horizontal limb.

- Whenever the **common bile duct (CBD) is incised**, it is sutured after inserting the T-tube. The short horizontal limb is placed vertically within the common bile duct after making 2–3 holes within. Some surgeons slit open the entire length of the short limb.
- The long limb is brought to the exterior from the most dependent part of the common bile duct and connected to a sterile container.
- Presence of the T-tube may prevent peritonitis due to biliary leakage in cases of residual stones blocking the lower end of the CBD.

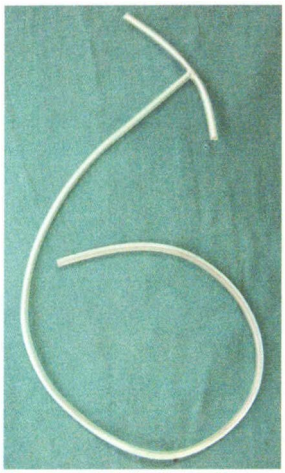


Fig. 51.57: T-tube (Kehr's)

REMOVAL OF THE TUBE

About 7–10 days later, a T-tube cholangiography is done and the T-tube is removed with a gentle pull, provided following criteria are fulfilled.

1. The dye flows freely into the duodenum.
2. No filling defects in the CBD.
3. After clamping the tube for 24 hours, there is no abdominal pain or fever.
4. Patient is passing normal coloured stools.

Once the tube is withdrawn, some amount of biliary leak may persist for 2–3 days and it stops by itself provided there is no distal obstruction.

SENGSTAKEN-BLAKEMORE DOUBLE BALLOON TRIPLE LUMEN TUBE (Fig. 51.58)

- It is used in controlling bleeding **oesophageal varices.** It has 3 lumens and 2 balloons, a gastric balloon and an oesophageal balloon.
- **Gastric balloon is inflated** with about **200–250 ml** of air and oesophageal balloon is inflated with about **40–60 ml of air**. It is pulled upwards so as to snugly fit at the oesophagogastric junction and thus it acts by internal tamponade.
- Sengstaken tube should not be kept in place for more than 48 hours because it can cause pressure necrosis of oesophagus.
- It should be deflated for a few minutes after 24 hours.

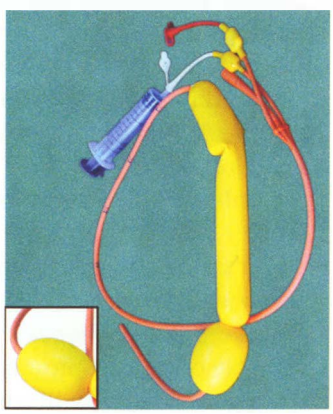


Fig. 51.58: Sengstaken-Blakemore double balloon triple lumen tube

- Sengstaken tube should be used by an experienced physician. Oesophageal secretions and saliva cannot be aspirated while using this tube, and if gastric balloon is deflated suddenly, it slides up and causes choking. The oesophageal balloon should be immediately deflated in such situations.
- **Modification of Sengstaken tube is called Minnesota tube or 4 lumen tube.** It has 4 lumens. The 1st to inflate oesophageal balloon, the 2nd to inflate gastric balloon, the 3rd to aspirate like a Ryle's tube, and the 4th lumen is used to aspirate oesophageal secretions. If there is any difficulty in breathing while using Sengstaken tube or Minnesota tube, bulb should be deflated or tube should be cut.

SUTURING NEEDLES (Fig. 51.59)

Traumatic

- Round body needle is an eyed needle. They are used to suture soft tissues, muscles, tendons, vessels, intestines, etc.
- Cutting needles are used to suture slim and some tough structures.
- Reverse cutting needle are used to suture mucoperiosteum.

These needles have an eye. The eye is wider than body of the needle, so tissue trauma is more.

Atraumatic

These needles have no eye. **Suture** is attached to the needle by a process called **swaging**. Tissue trauma is less, and hence is used in suturing vessels or to repair a small tear in the bowl, etc.

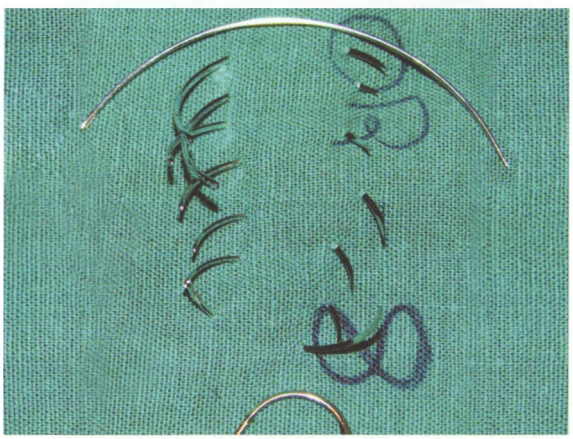


Fig. 51.59: Suturing needles

SUTURE MATERIALS (Key Box 51.3)

ABSORBABLE

1. Plain catgut (7-day catgut)

- The word catgut is derived from kit-gut, which means the violin strings. It is the oldest suture material known.
- Catgut is derived from the submucosa of the sheep intestines.

- The plain catgut lasts for **7–10 days**. Hence, its uses are minimal.
- It can be used to put 'fat stitches' (subcutaneous fat).
- It is biological, absorbable and monofilament.
- Sheep's submucosa has a rich content of elastic tissue.

2. Chromic catgut (21-day catgut)

- When plain catgut is mixed with chromic salts, chromic catgut is obtained.
- **The strength of the chromic catgut is about 15–25 days**.
- Chromic catgut is widely used in intestinal anastomosis, closure of urinary bladder, closure of common bile duct, gastrojejunostomy, etc.
- Catgut is biological, absorbable, monofilament suture material.
- Chromic catgut is packed along with round body needle.
- The number 2–0 refers to the thickness of the suture.
- Knotting property is good
- The catgut is preserved in 70% alcohol and is kept soft due to 5% glycerine.

3. Vicryl (polyglactin)

- This is a copolymer of glycolide and lactide.
- It is a synthetic and absorbable suture.
- Unlike catgut, this is absorbed by hydrolysis.
- **Since the strength and reliability is more than catgut, vicryl is being used more and more for small intestinal and colonic anastomosis. It has replaced catgut in suturing bile duct also.**
- Being synthetic, tissue reaction is less than that of chromic catgut.
- This has also replaced catgut while suturing common bile duct.
- Knotting property is good
- Vicryl can be used in the presence of infection.

4. Dexon (polyglycolic acid)

- Synthetic absorbable
- Braided
- Used like vicryl

KEY BOX 51.3

COLOUR	SUTURE MATERIAL
• Yellow	Plain catgut
• Brown	Chromic catgut
• Violet	Vicryl
• Creamy yellow	Dexon
• Creamy	PDS
• Blue	Prolene
• Black	Silk
• White	Cotton

5. PDS (polydioxanone suture)

- Like vicryl
- Costly
- Creamy in colour

NONABSORBABLE

1. Prolene

- This is polypropylene and nonabsorbable.
- It is monofilament, artificial and uncoated. Does nc harbour micro-organism. Hence, the chances of infectio are less.
- Since it is nonabsorbable, prolene can be used fo abdominal closure, repair of hernias, repair of incisiona hernia, etc.
- It has high memory (recoiling tendency after remova from the pocket) and hence multiple knots are required

2. Sutupack

- It is a monofilament or multifilament polyamide.
- Black in colour
- It is braided, uncoated and nonabsorbable.
- Uses of sutupack are similar to prolene.
- Knotting property is not very good. Hence, it i mandatory to put 4–5 knots.

3. Mersilk

- This is nonabsorbable, braided silk, black in colour.
- It has been provided with a round body. This can be usec in ligating bleeding points or anastomosis, etc.

4. Black silk

- This is a nonabsorbable suture material.
- It is biological and derived from the **cocoon of the silkworm larva**.
- It is braided, coated with wax to reduce capillary action. Tissue reaction is much more with black silk because it is a foreign protein.
- In spite of this, it is widely used because of its easy availability and is cheap.
- Knotting property is excellent

5. Cotton

- White in colour
- Multifilament—infection rate is high
- Nonabsorbable, cheap

WHAT IS NEW IN THIS CHAPTER?/RECENT ADVANCES

- All topics have been updated
- New photographs have been added

Note to students: *For more details about the instruments, you are requested to refer Manipal Manual of Instruments.*

52 Specimens

- TB lymphadenitis
- Hodgkin's lymphoma
- Carcinoma tongue
- Chronic gastric ulcer
- Linitis plastica
- Intussusception
- Carcinoma rectum
- Gangrenous appendicitis
- Carcinoma colon
- Meckel's diverticulum
- Polycystic kidney
- Renal cell carcinoma
- Hydronephrosis
- Carcinoma penis
- Seminoma testis
- Cholecystectomy
- Hydatid cyst
- Carcinoma stomach
- Lipoma
- Malignant melanoma
- Thyroidectomy
- Wide excision of skin

I. TUBERCULOUS (TB) LYMPHADENITIS

1. What is this specimen?
- Specimen of lymph nodes which are matted. Cut surface shows caseation. Hence, it is tuberculous lymphadenitis.

2. What is the microscopic picture?
- Central caseation surrounded by epithelioid cells, Langhans' type of giant cells.

3. What are the stages of TB lymphadenitis?
- Stage of lymphadenitis
- Stage of matting
- Stage of cold abscess
- Stage of collar stud abscess
- Stage of sinus formation

4. Why is matting seen in TB lymphadenitis?
- It is because of periadenitis

5. What is the treatment of cold abscess?
- Nondependent aspiration by using wide bore needle, t avoid sinus formation.

II. LYMPHOMA

1. What is the diagnosis?
- Multiple lymph nodes which are discrete and not matted Cut surface does not show caseation. It is homogenous Hence, this is a specimen of Hodgkin's lymphoma.

2. How do you confirm the diagnosis?
- Lymph node biopsy

3. What is the microscopic picture?
- Cellular pleomorphism: Lymphocytes, histiocytes eosinophils, monocytes with giant cells containing mirro image nuclei—Reed-Sternberg cell.

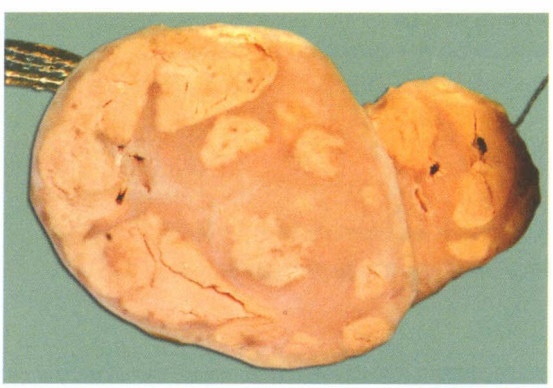


Fig. 52.1: Tuberculous (TB) lymphadenitis

Fig. 52.2: Lymphoma

4. **What are the common lymph nodes involved in Hodgkin's lymphoma?**
   - Cervical, axillary, para-aortic, iliac and inguinal lymph nodes.
5. **Is Waldeyer's ring involvement seen in Hodgkin's lymphoma?**
   - No, it is usually seen in non-Hodgkin's lymphoma.

III. SPECIMEN OF HEMIGLOSSECTOMY WITH HEMIMANDIBULECTOMY

1. **What is this specimen?**
   - Specimen showing growth arising from the tongue and infiltrating the mandible.
2. **What is the diagnosis?**
   - Advanced carcinoma tongue
3. **Is radiotherapy indicated in this situation?**
   - No, because chances of radionecrosis of the mandible are high.
4. **What type of X-ray is taken to look for involvement of the mandible?**
   - Orthopantomogram
5. **What is Commando's operation?**
   - Hemiglossectomy with excision of the floor of the mouth, hemimandibulectomy, with radical block dissection of the neck done in a single stage, with *en bloc* removal.

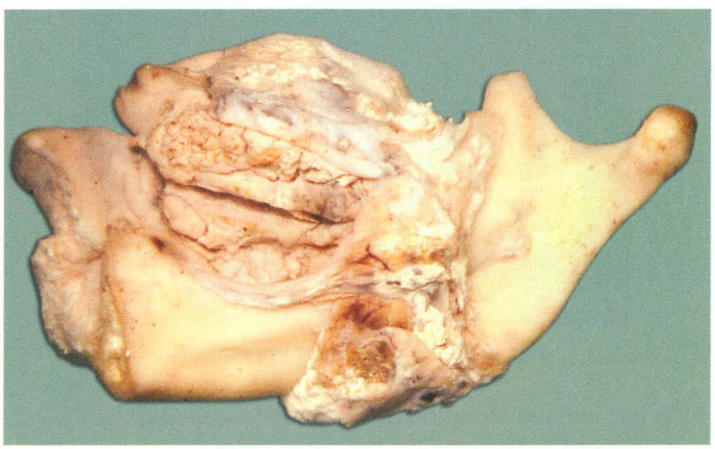


Fig. 52.3: Specimen of hemiglossectomy with hemimandibulectomy

IV. CHRONIC GASTRIC ULCER

1. **What is this specimen?**
   - Specimen of the stomach showing rugosity of the stomach. There is a deep ulcer crater along the lesser curvature.
2. **What is the diagnosis?**
   - Benign gastric ulcer
3. **Why is it a benign gastric ulcer?**
   - Since the rugae are of converging type, it is a benign gastric ulcer.
4. **How do you rule out malignancy in a gastric ulcer?**
   - Endoscopic biopsy

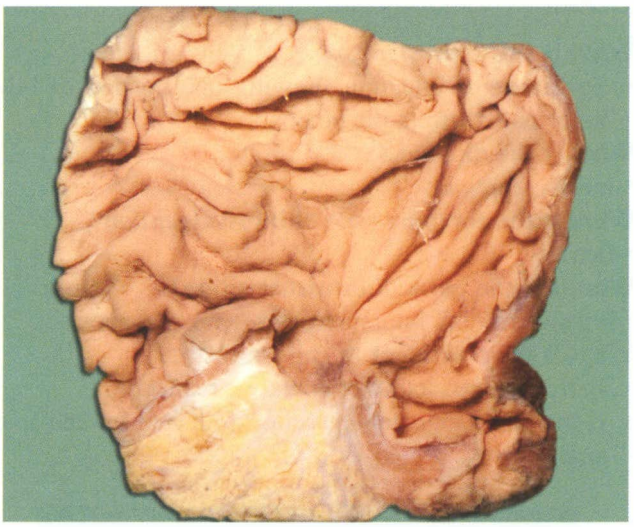


Fig. 52.4: Chronic gastric ulcer

5. **What is the incidence of gastric ulcer turning in malignancy?**
   - 0.5 to 2%

V. LINITIS PLASTICA

1. **What is this specimen?**
   - Specimen of the stomach showing loss of normal rugosi There is a nodular extensive infiltrating lesion along t entire length of the stomach.
2. **What is the diagnosis?**
   - Linitis plastica—leather bottle stomach.
3. **What is linitis plastica?**
   - It is an extensive fibrosis involving entire submucosa the stomach initially and involves other layers also late
4. **What is the treatment for linitis plastica?**
   - Radical total gastrectomy
5. **What is the prognosis?**
   - Very poor

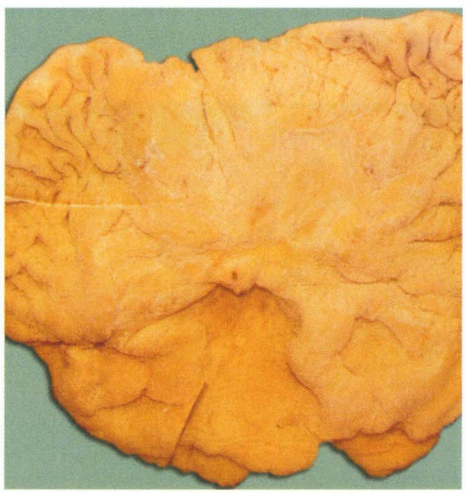


Fig. 52.5: Linitis plastica

VI. INTUSSUSCEPTION

1. What is this specimen?
- Specimen of intestine showing one portion of bowel invaginated within the other.

2. What is the diagnosis?
- Intussusception

3. What is the common type of intussusception?
- Ileocolic

4. What are the parts of intussusception?
- Intussusceptum, intussuscipiens, neck and apex.

5. What is the treatment in children?
- Hydrostatic reduction or operative reduction.
- If there is gangrene—resection followed by end-to-end anastomosis.

Fig. 52.6: Intussusception

VII. CARCINOMA RECTUM

1. What is this specimen?
- Specimen of rectum showing ulceroproliferative growth in the middle of the rectum. Specimen also shows entire rectum and anal canal.

2. What is the diagnosis?
- Carcinoma rectum

3. What is this surgery?
- Abdominoperineal resection (excision) (APR). In this operation, entire rectum, anal canal, part of the sigmoid colon, fat, fascia, lymphatics and regional nodes are removed *en bloc* followed by permanent colostomy in th left iliac fossa.

4. What are the indications for APR?
- Growth in the middle and lower rectum wherein sphincte cannot be saved.

5. What is the position of the patient during APR?
- Supine with lithotomy called Lloyd Davis position.

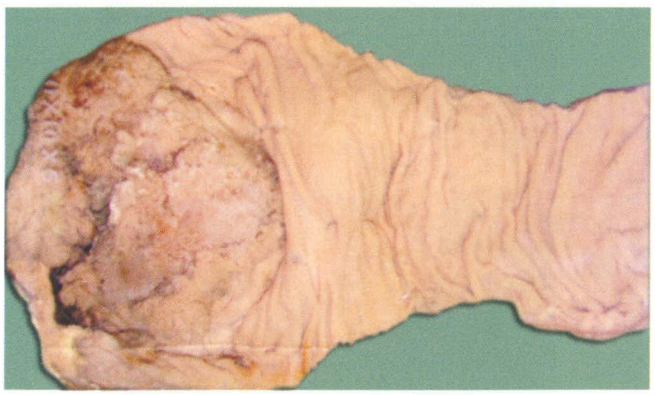


Fig. 52.7: Carcinoma rectum

VIII. GANGRENOUS APPENDICITIS

1. What is this specimen?
- It is an appendicectomy specimen showing blackis discolouration of the appendix.

2. What factors cause gangrene of the appendix?
- Gangrenous appendicitis occurs usually in elderly patients where there is decreased vascularity due to atherosclerosis It can also occur when the lumen is blocked due t faecolith, thereby causing ischaemia.

3. What is the one simple investigation which is useful i diagnosing appendicitis?
- Total WBC count. Above 10,000 cells/cu mm of bloo with increased neutrophil count.

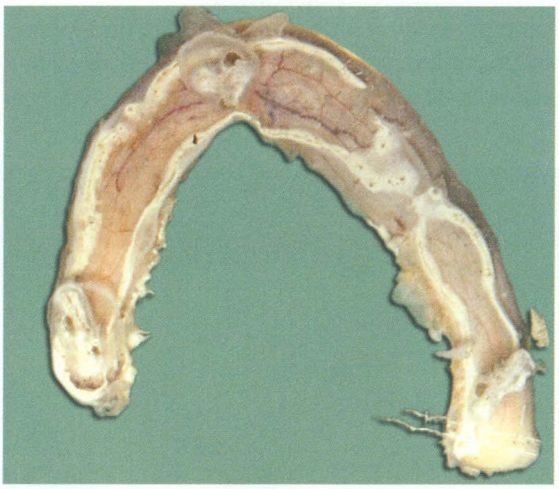


Fig. 52.8: Gangrenous appendicitis

4. What are the complications of acute appendicitis?
- Appendicular mass (in untreated cases)
- Perforation with an abscess
- Perforation with generalised peritonitis
- Pylephlebitis, portal pyaemia
- Septicaemia, gram-negative shock

5. How do you treat an appendicular mass?
- Conservative line, Oschner-Sherren regime—aspiration, antibiotics, intravenous fluids, etc.

IX. CARCINOMA ASCENDING COLON

1. What is this specimen?
- Specimen of terminal ileum, caecum and right colon with removal of involved lymph nodes and fat fascia. Nowadays, only 4–6 cm of ileum is removed.

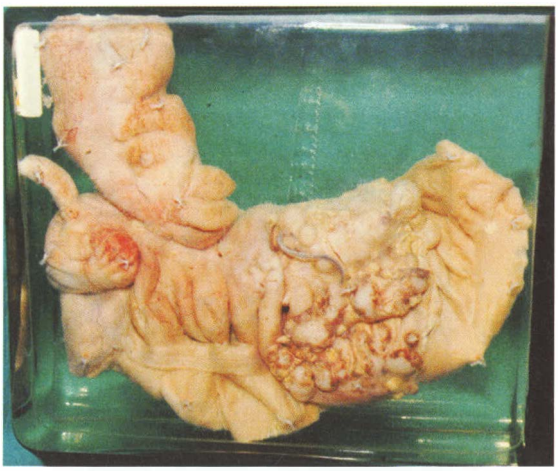


Fig. 52.9: Carcinoma ascending colon

2. What is the surgery?
- Right radical hemicolectomy done for growth in the ascending colon.

3. How do you identify colon?
- *Taenia coli* and appendix are seen. The colon has a larger diameter compared to small intestine.

4. What are the investigations?
- Barium enema will show persistent filling defect. However, colonoscopy is the investigation because the growth can be visualised and biopsy can be taken.

5. What do you mean by limited resection?
- It is done for ileocaecal tuberculosis wherein diseased segment is removed.

X. MECKEL'S DIVERTICULUM

1. What is the specimen?
- Resected specimen of intestine showing a diverticulum. Hence, it is a Meckel's diverticulum.

2. Why is it a Meckel's diverticulum?
- Because it is a single diverticulum arising from antimesenteric border of the intestine.

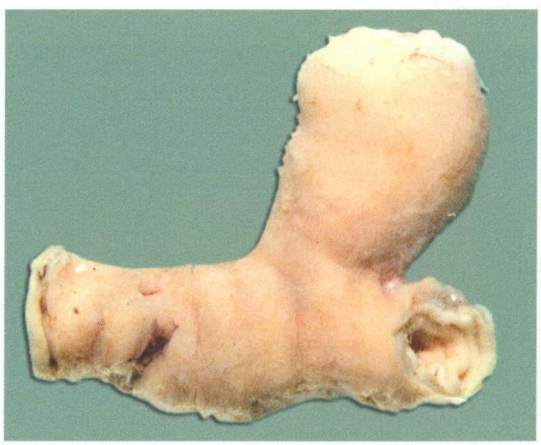


Fig. 52.10: Meckel's diverticulum

3. What are common symptoms?
- Bleeding per rectum, abdominal pain due to inflammatic intestinal obstruction and peritonitis due to perforatior

4. What is the cause and what are the types of bleeding?
- Ulcer in the ectopic gastric mucosa. Bleeding can occult, in small quantities or rarely can be massive.

5. How do you diagnose Meckel's diverticulum?
- Radio-nuclear (^{99m}Tc pertechnetate) scan is helpful wh there is active bleeding.

XI. POLYCYSTIC KIDNEY

1. What is this specimen?
- Specimen of kidney with multiple cystic lesions. Ent kidney is involved.

2. What is the diagnosis?
- Polycystic kidney

3. Why do you say it is polycystic kidney?
- Kidney is grossly enlarged
- Outer surface is bosselated
- Multiple cysts are present

4. What are clinical features of polycystic kidney?
- Women: 30–50 years
- Bilateral renal mass
- Hypertension
- Haematuria
- Renal failure

5. What is the treatment?
- If there is no renal failure, control hypertension.
- If there is renal failure—dialysis followed by ren transplantation.

Fig. 52.11: Polycystic kidney

XII. RENAL CELL CARCINOMA

1. What is this specimen?
- Specimen of the kidney because it is reniform shaped, ureter and calyces are seen.
- In the upper pole, there is destruction of the calyces with solid mass. Cut surface is smooth.

2. What is the diagnosis?
- Renal cell carcinoma

3. What is the microscopic picture?
- Cuboidal or polyhedral clear cells with deeply stained rounded nuclei—clear cell carcinoma. Sometimes, dark cells can coexist. In some cases, walls of blood vessels are lined by tumour cells.

4. How does it spread?
- Lymphatic and blood spread

5. What are the primary malignant tumours which spread by blood?
- Renal cell carcinoma, follicular carcinoma thyroid, carcinoma prostate, carcinoma breast, bronchogenic carcinoma.

Fig. 52.12: Renal cell carcinoma

XIII. HYDRONEPHROSIS

1. What is this specimen?
- Specimen of the kidney with ureter showing dilatation of pelvicalyceal system. Calyces are club-shaped.

2. What is the diagnosis?
- Hydronephrosis—probably due to pelvi-ureteric junction (PUJ) obstruction.

3. Why PUJ obstruction?
- Ureter is not dilated

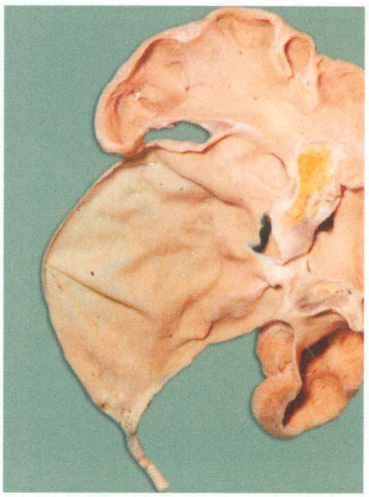


Fig. 52.13: Hydronephrosis

4. What are the common causes of obstruction at PUJ?
- Stone in the pelvis
- Aberrant vessels—a lower polar artery or vein arising from the main vessels in an aberrant position obstructs the upper ureter.
- PUJ dyskinesia—occurs due to incoordination between neuromuscular impulses and pelvis.

5. What is the treatment of PUJ dyskinesia?
- Anderson-Hynes pyeloplasty.

XIV. CARCINOMA PENIS

1. What is this specimen?
- Specimen of penis, showing the glans. Prepuce is cut open showing the growth.

2. What is the diagnosis?
- Partial amputation done for carcinoma penis

3. What are the indications for partial amputation of the penis?
- Growth confined to the glans penis or to the prepuce.

4. If shaft is involved, what is the treatment?
- Total amputation of penis followed by perineal urethrostomy.

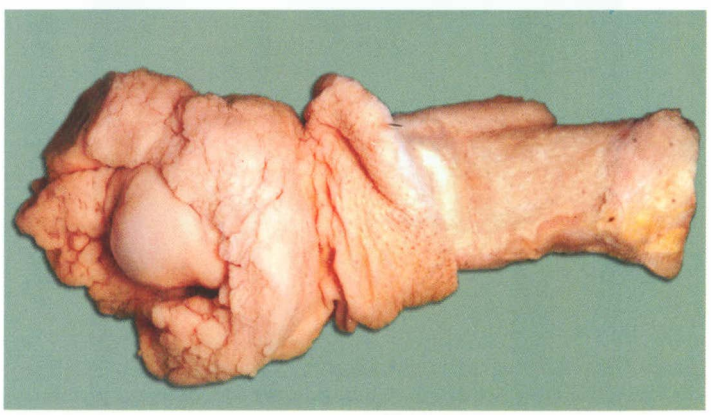


Fig. 52.14: Carcinoma penis

5. What are the complications of perineal urethrostomy?
- Bleeding, dermatitis and stenosis. The stenosis should be dilated by using Hegar's dilators.

XV. SEMINOMA TESTIS

1. What is this specimen?
- Specimen of testis showing spermatic cord. Cut surface of the testis is smooth and homogenous with a tumour in the upper part.

2. What is the diagnosis?
- Seminoma

3. Why not a teratoma?
- In a teratoma, the cut surface is not homogenous.

4. How does seminoma spread?
- Mainly by lymphatics

5. What type of orchidectomy is done for testicular tumours and why?
- High orchidectomy, through an inguinal incision. If scrotum is incised, chances of alternate pathway of lymphatics opening up are high. Hence, inguinal exploration is the choice.

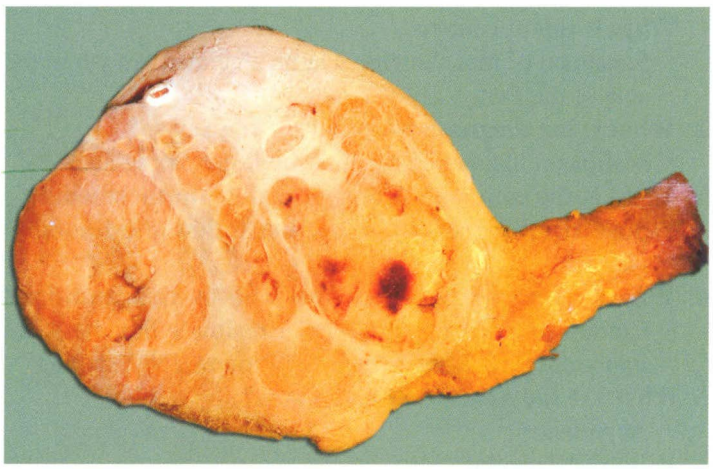


Fig. 52.15: Seminoma testis showing spermatic cord on the right side

XVI. CHOLECYSTECTOMY FOR GALL STONES

1. What is this specimen?
- Cholecystectomy specimen

2. What is the diagnosis?
- Multiple stones are present within lumen—diagnosis is probably chronic cholecystitis.

3. Why do you say it is a gall bladder?
- It is pear-shaped with fundus, body and a narrow portion—cystic area.

4. What is Hartmann's pouch?
- It is the distal angulated portion of gall bladder wherein a stone commonly lodges.

5. What are the common symptoms of gall stones?
- Flatulent dyspepsia, gall stone colic, acute and chroi cholecystitis are common symptoms of gall ston Mucocoele, empyema, perforation and gall sto pancreatitis are other complications.

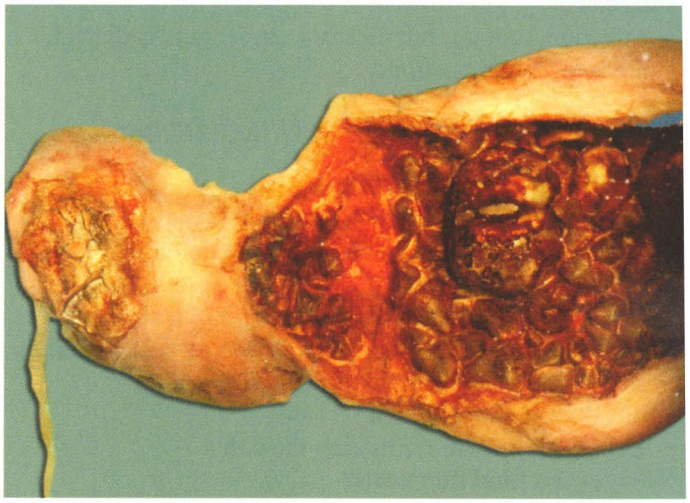


Fig. 52.16: Gall stones

XVII. HYDATID CYST

1. What is this specimen?
- Specimen of laminated membranes—this layer is als called ectocyst. It is thick and elastic resembling onic skin appearance.

2. What is the diagnosis?
- Hydatid cyst—mostly liver

3. What are the other layers of hydatid cyst?
- Outermost layer is called adventitial layer which blenc firmly with liver tissue. The middle layer is ectocyst, als called laminated membrane. Inner layer is germin epithelium, also called endocyst within which broo capsules and daughter cysts are present.

Fig. 52.17: Hydatid cyst

4. **What is the drug for hydatid disease?**
   - Albendazole 400 mg, a day for 15 days followed by no drug for 15 days. Then restart the cycle. Such treatment may have to continue for 6 months depending on the response rate.
5. **What are the common complications of hydatid cyst of the liver?**
   - Infection, rupture, calcification, cholangitis with jaundice are a few complications.

XVIII. RADICAL GASTRECTOMY INCLUDING REMOVAL OF THE COLON

1. **What is the specimen?**
   - Specimen of stomach with transverse colon.
2. **Why stomach and colon?**
   - It has lesser curvature and greater curvature—pylorus, body and proximal stomach. Colon is the immediate structure below the stomach.
3. **What does it show?**
   - Exophytic growth infiltrating the colon
4. **What is the final diagnosis?**
   - Most probably it is carcinoma stomach infiltrating transverse colon.

Fig. 52.18: Radical gastrectomy including removal of the colon

5. **What is the best investigation in such cases to identify local infiltration?**
   - CT scan

XIX. LIPOMA

1. **What is this specimen?**
   - Specimen of lipoma
2. **Why do you say it is lipoma?**
   - It is lobular, yellow in colour
3. **What is the commonest site and type of lipoma?**
   - Flank is the commonest site. Single and subcutaneous variety is the commonest type.

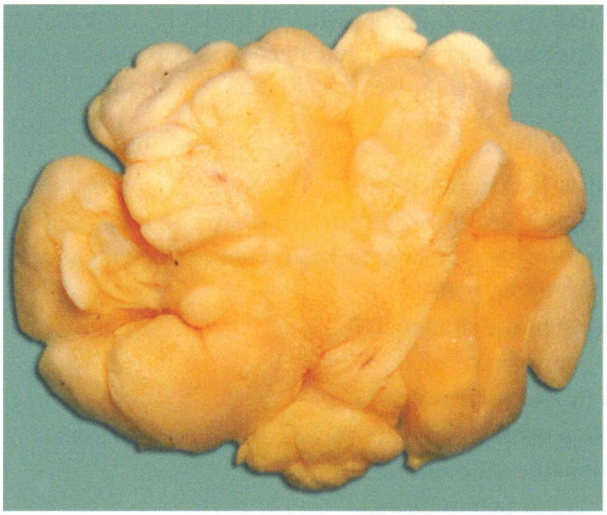


Fig. 52.19: Lipoma

4. **What are the common complications of lipoma?**
   - Liposarcoma and intussusception
5. **Which type of lipoma give rise to intussusception?**
   - Submucosal type

XX. MALIGNANT MELANOMA

1. **What is this specimen?**
   - Specimen of foot showing a large ulcerated growth in th sole of the foot.
2. **What is the diagnosis and why do you say so?**
   - Malignant melanoma because the lesion is pigmented.
3. **What is the commonest type of malignant melanoma?**
   - Superficial spreading is the first followed by nodula variety.
4. **What are the staging systems available for this condition**
   - Clark's level of invasion and Breslow's thickness ar important staging systems in addition to TNM staging.
5. **What are the ABCDE of melanoma?**
   - Asymmetry
   - Border irregular
   - Colour variegation
   - Diameter > 6 mm
   - Elevation

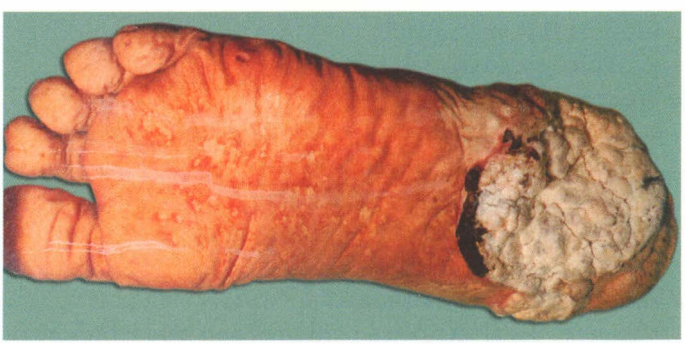


Fig. 52.20: Malignant melanoma

XXI. THYROIDECTOMY SPECIMEN

1. **What is this specimen?**
   - Specimen of thyroid gland showing both lobes and isthmus.
2. **What is the diagnosis and why do you say so?**
   - Probably it is a subtotal thyroidectomy specimen— surgery is done for multinodular goitre.
3. **What is the commonest type of malignancy of the thyroid gland?**
   - Papillary carcinoma—63%. 2nd common type is follicular carcinoma thyroid.
4. **What is the surgical treatment for well-differentiated carcinoma thyroid gland?**
   - Most centres follow total thyroidectomy. If the patient is in low-risk category, lobectomy can be done.
5. **What blood investigation is useful in the follow-up period of papillary carcinoma thyroid gland?**
   - Thyroglobulin

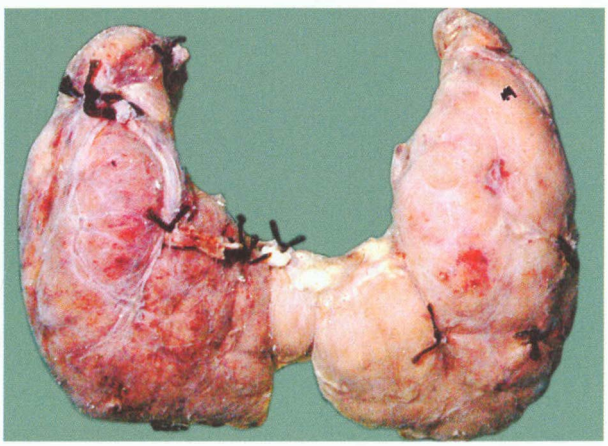


Fig. 52.21: Thyroidectomy specimen

XXII. WIDE EXCISION SPECIMEN OF SKIN

1. **What is this specimen?**
   - Specimen of skin which has been excised with normal skin. Hence, wide excision specimen.
2. **What is the diagnosis and why do you say so?**
   - Probably it is a squamous cell carcinoma because edges are everted.
3. **What are the common sites of squamous cell carcinoma skin?**
   - Areas with chronic irritation, e.g. Kangri cancer in the abdominal wall, chimney sweepers cancer, etc.
4. **What are the common precancerous lesions for squamous cell carcinoma?**
   - Leukoplakia, Bowen's disease, chronic irritation and scar tissues, etc.
5. **What do you call squamous cell carcinoma arising in a scar tissue?**
   - Marjolin's ulcer

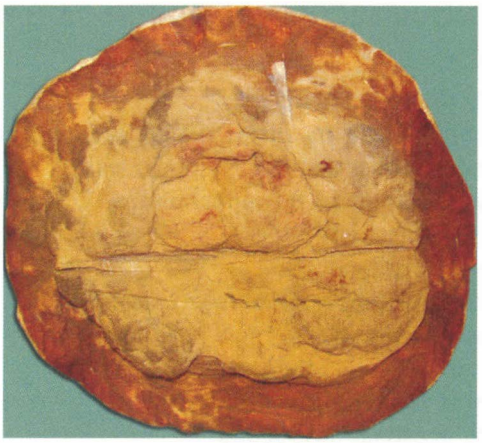


Fig. 52.22: Wide excision skin

XXIII. WHIPPLE'S PANCREATICODUODENECTOMY

1. **What is this specimen?**
   - Specimen showing distal stomach, duodenum, proxim jejunum and pancreatic head.
2. **What is the name of this operation?**
   - Whipple's pancreaticoduodenectomy
3. **Why is it done?**
   - Mostly due to periampullary carcinoma
4. **How can you get histopathological diagnosis?**
   - Endoscopic biopsy
5. **Is there any other indication for Whipple's surgery?**
   - Pancreatic head mass—doubt exists between chron pancreatitis and carcinoma head pancreas. Provide experience of the surgeon is good.

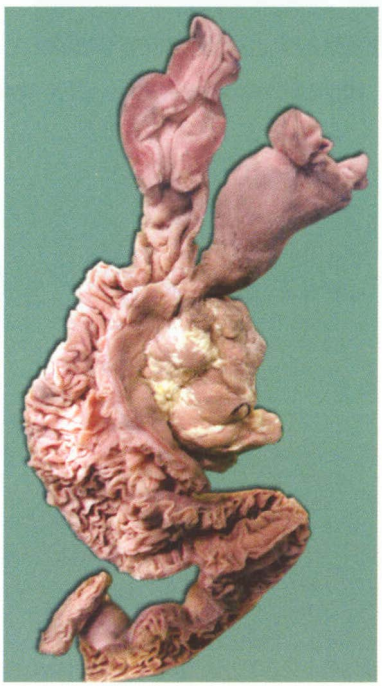


Fig. 52.23: Whipple's pancreaticoduodenectomy

XXIV. RIGHT HEMICOLECTOMY FOR CARCINOMA CAECUM

1. **What is this specimen?**
   - Specimen showing distal ileum, caecum, part of the ascending colon and few lymph nodes
2. **What is the name of this operation?**
   - Limited colectomy
3. **Why is it done?**
   - Mostly due to ileo-caecal tuberculosis
4. **Why do you say so?**
   - Stricture is seen in the terminal ileum
5. **What was the indication for surgery?**
   - Acute intestinal obstruction

Fig. 52.24: Right hemicolectomy for carcinoma caecum

XXV. SPLENECTOMY SPECIMEN

1. **What is this specimen?**
   - Specimen showing spleen with laceration of tl diaphragmatic surface.
2. **Why laceration?**
   - Blunt injury is the most common cause of rupture splee
3. **What will be clinical manifestation?**
   - Bleeding
4. **What other surgery can be done for bleeding?**
   - Partial splenectomy or splenorrhaphy
5. **What is the dangerous complication after splenectomy**
   - Opportunistic post-splenectomy infections

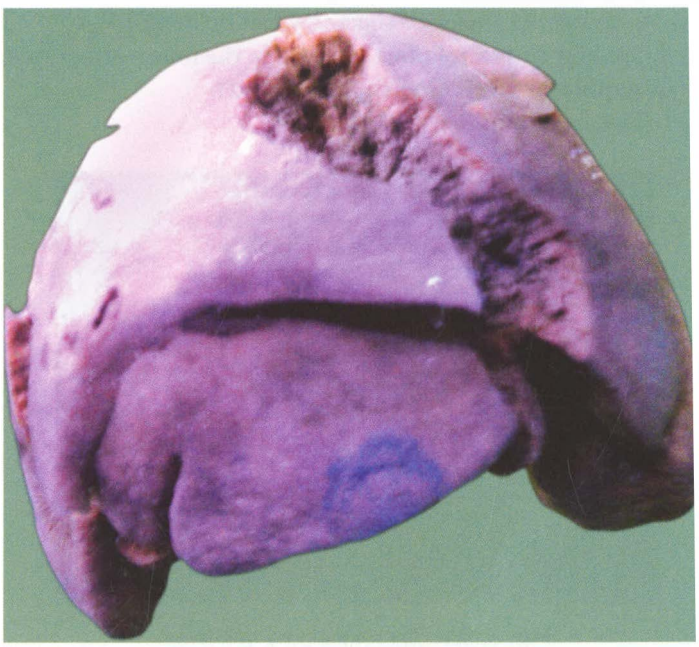


Fig. 52.25: Splenectomy specimen

Useful Tips

- Please look into the specimen carefully
- Please see both sides of the specimen
- Think which is the most likely organ involved
- Think what is the probable diagnosis.

53

Operative Surgery, Laparoscopic Surgery and Accessories

- Appendicectomy
- Herniorrhaphy: Bassini
- Surgery for hydrocoele
- Incision and drainage (I & D)
- Incision and drainage of breast abscess
- Circumcision
- Venesection or cut down
- Vasectomy
- Tracheostomy
- Suprapubic cystostomy (SPC)
- Thyroidectomy
- Amputations
- Amputations in leg
- Upper limb amputations
- Open cholecystectomy
- Vagotomy gastrojejunostomy (GJ)
- Excision of swellings
- Colonic surgery
- Laparoscopic surgery
- High frequency (HF) electrosurgery
- Cryosurgery
- Lasers in surgery
- Staplers in surgery

Introduction

Over a period of time, the number of operations an undergraduate is expected to know has become less and less. Today no MBBS doctor is supposed to do a surgical procedure because qualified surgeons are available even in a village. Hence, I have discussed a few surgical procedures in this chapter. These are common surgical procedures that an undergraduate student is expected to know. Every operation has been discussed along a certain basic pattern as given below and the key words used are given in the key box. *Students should study surgical anatomy before reading this chapter.*

Commonly used abbreviations (Key Box 53.1)

Steps of operative surgery

1. Indications
2. Contraindications
3. Position of the patient
4. Anaesthesia
5. Preparation of parts
6. Procedure
7. Closure
8. Postoperative management
9. Postoperative complications
10. Advice at discharge

KEY BOX 53.1

COMMONLY USED ABBREVIATIONS

SA	Spinal anaesthesia
GA	General anaesthesia
LA	Local anaesthesia
OT	Operation theatre
NPO	Nil per oral
IV	Intravenous
RT	Ryle's tube

ANTISEPTIC AGENTS

- Povidone-iodine
- Spirit 70%
- Savlon

Autoclave and sterilisation (kindly *see* Table 53.1 in next page and page 3 of *Manipal Manual of Surgery for Dental Students*, 3rd edition for details)

APPENDICECTOMY

PEARLS OF WISDOM

Appendicectomy can be one of the easiest and sometimes one of the most complicated surgeries.

1. Indications

- Acute appendicitis—emergency appendicectomy
- Recurrent appendicitis—elective appendicectomy

Table 53.1 Sterilisation

Agents for sterilisation	Common of items
1. Autoclaving	Linen, operative instruments, glass syringes
2. Dettol or phenol	Sharp instruments (scissors, needles, blades)
3. Glutaraldehyde	Endoscopy and laparoscopy equipment
4. Ethylene oxide gamma radiation	Surgical catgut, syringes
5. Formaldehyde	Disinfect rooms like OT
6. Skin	70% spirit, povidone-iodine

2. Contraindications

Appendicular mass

3. Position of the patient

Supine

4. Anaesthesia of the parts

This surgery can be done either under GA or regional anaesthesia (spinal or epidural).

5. Preparation

Parts are cleaned with iodine and spirit, from the level of umbilicus above to the upper part of thigh below.

6. Procedure

Incision

1. McBurney's grid-iron incision is the most popular incision. It is at right angles to spino-umbilical line placed at McBurney's point. It is about 6–8 cm in length (Fig. 53.1).
2. **Lanz** incision is a curved transverse incision, placed at the McBurney's point. Cosmetically, it is a better incision (Fig. 53.2).
3. Right paramedian incision is made when diagnosis is in doubt as a part of **exploratory laparotomy**. This is also preferred in females where there is a gynaecological pathology such as ovarian cyst which may be the cause of right iliac fossa pain (Fig. 53.3). ***Today, all such cases are managed by lower midline incision.***

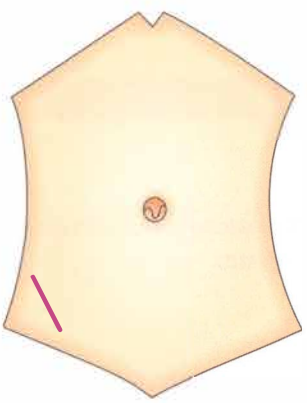


Fig. 53.1: Grid iron incision

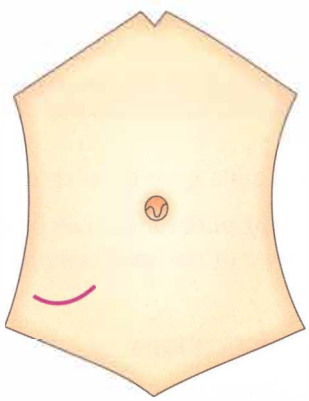


Fig. 53.2: Lanz incision

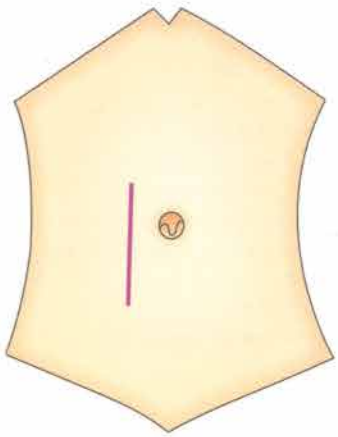


Fig. 53.3: Right paramedian incision

Layers opened

- Skin
- Two layers of subcutaneous tissue—superficial fatty (Camper's), deep membranous (Scarpa's). (C: comes first, S: later). There is no deep fascia in the abdomen.
- External oblique aponeurosis is seen running downwards and medially. It is incised in the direction of its fibres.
- Internal and transverse abdominal muscles are split (Grid Iron—right angle to each other).
- Peritoneum is incised.

Features of acute appendicitis at operation

- Inflamed, turgid appendix
- Pus in the right iliac fossa
- Presence of omentum in the right iliac fossa
- Black or green appendix (gangrenous)
- Faecolith

Location of appendix

- Trace taenia coli. They will lead to the base of appendix (**all roads lead to Rome**).
- When you are tracing taenia coli, if you are not able to identify the appendix, it means most probably you are tracing taenia coli of the sigmoid colon.

Surgical procedure

- Appendix is gently held at mesoappendix by using Babcock's forceps and blood vessels in the mesoappendix are divided. These include appendicular artery, branch of ileocolic artery (accessory appendicular artery of Seshachalam, is a branch of posterior caecal artery). Once appendix is freed up to the base (caecum), a **purse string suture** is applied all round appendix, taking bites from caecum, using 2–0 atraumatic silk (Fig. 53.4).

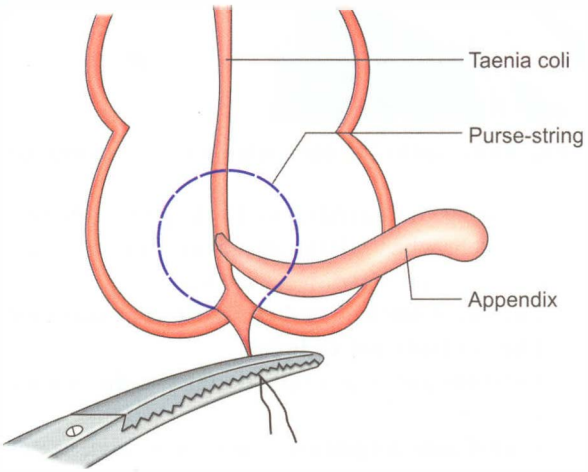


Fig. 53.4: Purse string suture

- Appendix is crushed at base and is held 1 cm above the crush. A tight silk ligature is applied at the crushed site and appendix is cut in between. Stump is cleaned with spirit, invaginated and purse string is tightened. This is called burial of the stump. Perfect haemostasis is obtained (Fig. 53.5).

PEARLS OF WISDOM

Look for Meckel's diverticulum, which may be the cause of right iliac fossa pain.

7. Closure

- Peritoneum—continuous 2–0 catgut/vicryl
- Split muscles—sutured together by a few interrupted sutures using chromic catgut/vicryl
- External oblique is sutured with silk
- Subcutaneous fat is sutured with vicryl
- Skin with interrupted silk
  Instead of chromic catgut, 2–0 silk, 2–0 vicryl is being used more often nowadays.
- Corrugated red rubber drain is not kept routinely unless there is gangrenous appendicitis or a lot of pus in the peritoneal cavity.

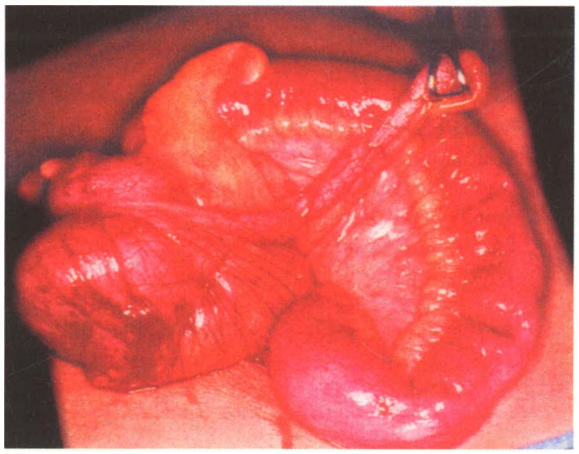


Fig. 53.5: Appendix at surgery

8. Postoperative management

- RT aspiration for one or two days.
- IV fluids 2.5 litres/day for one or two days.
- Oral fluids are allowed once abdomen is soft and bowel sounds are heard.
- Appropriate antibiotics to cover gram-positive, gram-negative and anaerobic organisms.
- Suture removal by 7–10 days.

9. Complications after appendicectomy

A. Postoperative fever can be due to various factors. Thrombophlebitis, urinary tract infection and IV fluids are common causes. In the absence of these, wound infection, intraperitoneal abscess secondary to gangrenous appendicitis, may have to be considered.
- Change of antibiotics according to culture and sensitivity reports of urine, pus and blood help in treating postoperative fever.
- Elderly patients may have a pre-existing pulmonary disease. Respiratory tract infection also has to be considered.

B. Wound infection: It is the most common complication after appendicectomy

C. Intra-abdominal abscess needs drainage

D. Faecal fistula—causes

a. Gangrene spreading into caecum
b. Persistent infection
c. Carcinoma caecum (elderly patients)
d. Ileocaecal tuberculosis
e. Crohn's disease (uncommon in India)
f. Actinomycosis (rare)[1]

PEARLS OF WISDOM

Most of the faecal fistulae will stop by themselves provided there is no distal obstruction.

[1]In viva, when a question is asked as to what is the common cause of faecal fistula following appendicectomy, the usual answer by students is actinomycosis. Remember it is the answer to be told last.

E. Septicaemia, portal pyaemia, gram-negative shock in late cases of peritonitis due to perforated appendicitis are uncommon but dangerous complications.

- Mortality of appendicular perforation and peritonitis is around 2%.

10. Advice at discharge

Rest for 15 days

HERNIORRHAPHY: BASSINI

- This means herniotomy and approximation of conjoined tendon to inguinal ligament to strengthen the posterior wall of the inguinal canal.
- In a large, long-standing hernias and in sliding hernias, especially in elderly patients, it is better to catheterise their bladder before surgery for two reasons. Firstly, to avoid injury to the urinary bladder and secondly, they invariably develop retention of urine in the postoperative period.

1. Indication

Indirect or direct hernia with good muscle tone

2. Contraindication (relative)

Severe cardiopulmonary insufficiency

3. Position of the patient

Supine

4. Anaesthesia

Regional anaesthesia or G/A. **Local anaesthesia** can be preferred in **high-risk patients**.

5. Preparation of the parts

Like that for appendicectomy

6. Procedure

Incision

6–8 cm incision is made parallel to the inguinal ligament at the level of deep ring in the medial two-thirds of the inguinal ligament.

Layers opened

- Skin
- Two layers of superficial fascia
- External oblique is incised in the line of direction of fibres, till external ring is slit open.
- Thin cremasteric box is opened
- Identification of the sac—glistening white colour.
  - Isolate the cord from the sac by blunt and sharp dissection. The cord is held separately by using cord holding forceps (Fig. 53.6).

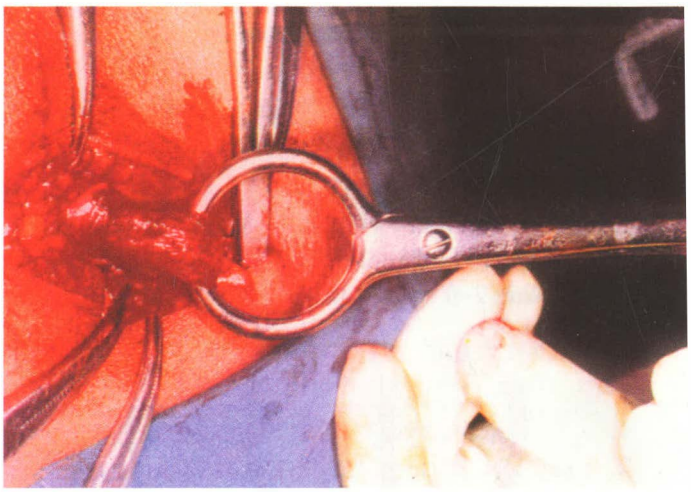


Fig. 53.6: Spermatic cord held with cord holding forceps

- The sac is mobilised up to the deep ring. Mobilisation is complete when inferior epigastric artery pulsations and extraperitoneal pad of fat are seen.
- **The sac is opened and contents are examined**
- **The contents are reduced**
- **Twist the sac** so as to avoid injury to the contents of the sac (Fig. 53.7).
- **Transfixation ligature** is applied as high as possible at the neck of sac and it is tightened.
- **Excision of the sac:** After excision, see the excised sac and see whether omentum or intestine have been injured. Up to this stage, it is called **herniotomy**.

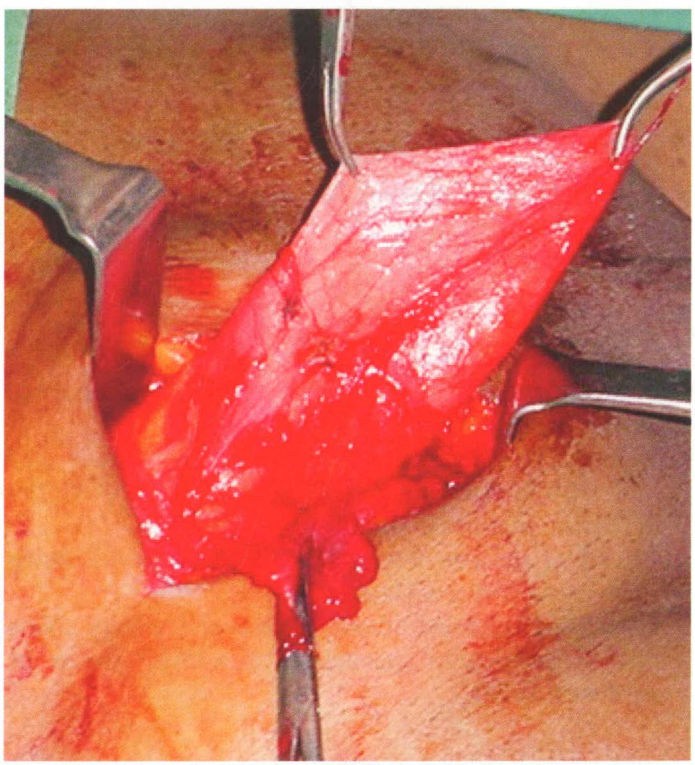


Fig. 53.7A: Direct hernial sac is usually not opened

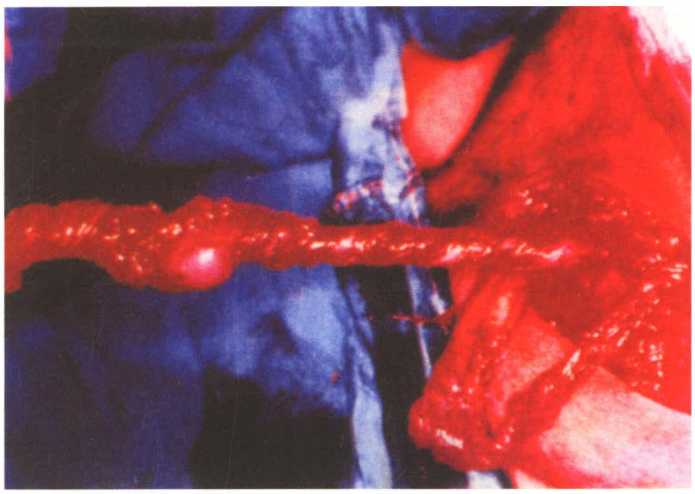


Fig. 53.7B: Twisted hernial sac ready for transfixation

REPAIR

- Conjoined tendon above is approximated to the inguinal ligament below by using nonabsorbable suture such as nylon, silk or sutupack.
- Nonabsorbable suture is used so that its strength remains for a long time. This repair is called as **Bassini's herniorrhaphy** (Key Box 53.2).

PEARLS OF WISDOM

Do not twist the sac in direct hernias and in sliding hernias.

KEY BOX 53.2

HERNIORRHAPHY

- Identify the sac
- Isolate the sac
- Mobilise the sac
- Open the sac
- Reduce the contents
- Twist the sac
- Transfixation ligation of the sac
- Repair

Precautions (Key Box 53.3)

1. Ilioinguinal nerve should not be caught in ligature.
2. Conjoined muscles should not be strangulated.
3. There should not be any tension in the suture lines.

7. Closure

- External oblique is sutured with chromic catgut or silk.
- Subcutaneous fat with absorbable catgut suture.
- Skin with silk.

8. Postoperative management

- NPO for 6–8 hours, oral fluids and soft diet later.
- Analgesics.
- Antibiotics—not always necessary.

KEY BOX 53.3

TAKE CARE OF

- 1 nerve—ilioinguinal nerve. Entrapment causes inguinal neuralgia (*see* page 843)
- 1 bone—pubic bone do not take a stitch from the bone. It may cause periostitis pubis
- 1 cord—spermatic cord do not tighten the deep ring too tight
- 1 small artery—inferior epigastric artery—see the pulsations near the deep ring and protect it
- 1 large artery—external iliac artery—while inguinal ligament being sutured to conjoined tendon on the lateral sides. Feel the pulse and protect
- 1 organ—urinary bladder—especially in elderly patients, large hernias and sliding hernias

- Scrotal support if the dissection is more (complete hernia).
- Suture removal after 7–10 days.

9. Postoperative complications

1. ***Immediate:*** Haematoma due to injury to the pampiniform plexus of veins or improper haemostasis. It may need re-exploration.
2. ***Wound infection*** may result in discharging pus which is the cause of postoperative fever. Infection is the chief cause of recurrence.
3. ***Severe periostitis pubis*** (to avoid this nowadays, the repair is not done by taking bites through pubic bone). X-ray of the bone may have to be taken for diagnosis.
   - It is managed by analgesics and in intractable cases, injection of corticosteroids locally may reduce the pain.
4. Nerve entrapment causing pain.

10. Advice at discharge

- Not to strain or lift heavy weights (e.g. bucketful of water) or to carry load on the shoulders for 3 months.
- If there is any precipitating cause such as chronic cough or difficulty in passing urine, etc. they have to be treated first. Otherwise, hernia will recur once again.

PEARLS OF WISDOM

Dissecting an inguinal canal and performing a good repair is a good exercise for a postgraduate because he has to dissect various anatomical layers, preserve nerves, vessels, vas deferens and do a good repair. Various steps of herniorrhaphy teach a postgraduate basic fundamental principles of surgery—Prof. C.R. Ballal, ex-Professor and Head, Kasturba Medical College and Hospital, Mangalore.

Today, the procedures of choice for inguinal hernia is mesh repair (*see* page 852)

SURGERY FOR HYDROCOELE

1. Indication

Vaginal hydrocoele. However, infantile and funicular hydrocoele are also treated surgically in the same manner.

2. Contraindication

Secondary hydrocoele due to *testicular tumours*. They contain haemorrhagic fluid. **Biggest blunder** can be done here by incising the scrotum mistaking it to be a vaginal hydrocoele.

3. Position of the patient

Supine

PEARLS OF WISDOM

Before incising scrotum, rule out testicular tumour. When in doubt, get ultrasound of testis.

4. Anaesthesia

- SA or GA.
- It can also be done by using local infiltration anaesthesia.

5. Preparation of the parts

Savlon and spirit (iodine is better avoided because it can cause severe scrotal dermatitis and excoriation of skin, which can cause more discomfort to the patient than hydrocoele surgery).

6. Procedure (Key Box 53.4)

Incision

Hydrocoele is held tense by an assistant and 5–6 cm incision (depending upon size) is made over the most prominent part of the swelling parallel to the median raphe of the scrotum.

Layers opened

- Skin
- Dartos
- External spermatic fascia
- Cremasteric fascia
- Internal spermatic fascia

At this stage hydrocoele sac is visible and is delivered outside the incision (Fig. 53.8).

Hydrocoele fluid is drained by using ***trocar and cannula.*** An opening is made in the tunica vaginalis sac and it is enlarged. All fluid is drained out. Testis and epididymis are inspected for any pathology, e.g. Craggy epididymis can found in tuberculosis. Depending on the size of the hydrocoe and thickness of the wall of the sac, two types of surgery c be done.

1. Small tunica vaginalis sac (TV sac)

- The redundant tunica vaginalis is plicated by interrupte sutures. The sac gets crumpled up and surrounds th testis. This is called **Lord's plication** (Fig. 53.9).

2. Sac is large and thick

- Partial excision and eversion of sac is ideal treatmen In this operation, after excision of the sac, cut edge c the sac is everted and sutured behind the testis. This i called **Jaboulay's operation**. **By eversion of the sac the secreting surface of the testis becomes anterio and secretions are absorbed by subcutaneou lymphatics** (Fig. 53.10A and 53.10B).

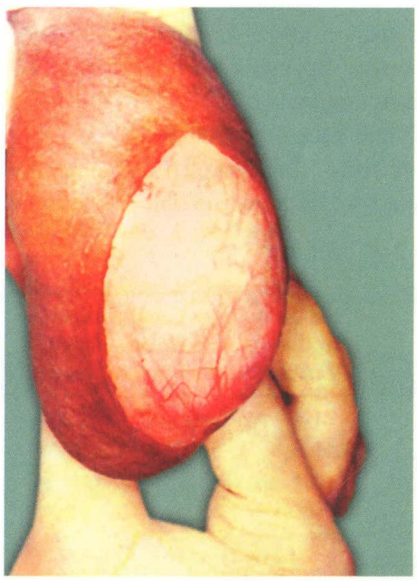


Fig. 53.8: Hydrocoele sac being delivered

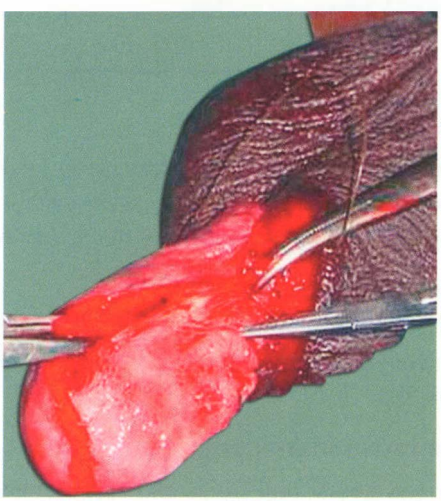
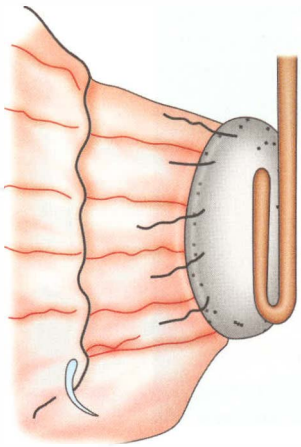


Fig. 53.9: The sac has been everted and edges of the sac sutured together

KEY BOX 53.4

HYDROCOELE SURGERY

- Aspiration: Not advised
- Lord's plication: Small hydrocoele
- Jaboulay's: Large hydrocoele



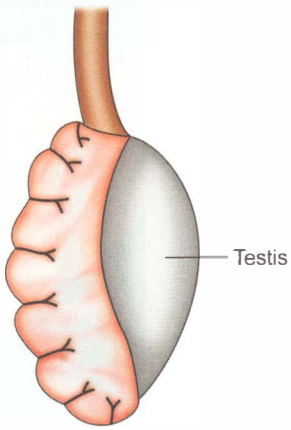


Fig. 53.10A: Lord's plication **Fig. 53.10B:** Jaboulay's operation

7. Closure

- A corrugated plastic drain is kept in the scrotum and brought out separately by making a stab incision and is anchored to the scrotal skin by white thread.
- Subcutaneous layer by using absorbable suture such as chromic catgut or vicryl sutures.
- Skin—interrupted thread (white)/catgut. Silk is avoided for skin closure over the scrotum as black colour of the silk is not seen clearly over dark pigmented skin of the scrotum, making stitch removal difficult. Absorbable sutures such as catgut or vicryl can also be used.
- Scrotal support is given to reduce oedema.

8. Postoperative management

- NPO for 6 hours followed by soft diet.
- Antibiotics and analgesics
- Suture removal after 7–8 days.

9. Postoperative complications

- **Haematoma:** If it is large and increasing, wound should be reopened urgently and bleeders have to be ligated. It may be due to injury to the testicular artery, vein or pampiniform plexus of veins.
- **Wound infection** can result in pyocoele. Testis can undergo necrosis. Such cases are treated with orchidectomy.
- **Injury to the spermatic cord**.

10. Advice at discharge

Rest for about a week

PEARLS OF WISDOM

Even though surgery for hydrocoele is minor, it should not be taken lightly.

INCISION AND DRAINAGE (I & D)

1. Indication

Pyogenic abscess

2. Contraindication

Cold abscess

3. Position of the patient

Supine, prone or lateral depending upon site of abscess.

4. Anaesthesia

- GA is preferred because abscess is multiloculated and infiltration of lignocaine into the abscess cavity does not act because of the acidic pH of the pus.
- However, a superficial abscess which is pointing can be managed without GA.

5. Preparation of the parts

Iodine and spirit.

6. Procedure

- **A stab incision** is made over the most prominent part of the swelling where skin is red, thinned out and is pointed.
- Pus that is drained is ***sent for culture and sensitivity.***
- A sinus forceps or finger is introduced within the abscess cavity and all the loculi are broken. When fresh blood oozes out, it indicates the completion of the procedure.
- Cavity is irrigated with antiseptic agents such as iodine solution. It is followed by irrigation with normal saline.
- If cavity is large, it is packed with roller gauze soaked in iodine and it is removed after 24–48 hours. Packing helps in controlling the bleeding, and keeps the abscess cavity open. By 7–10 days, the cavity collapses, granulation tissue fills up the cavity and healing takes place.

7. Closure

An abscess should not be closed, as it contains pus, bacteria (*See* also breast abscess drainage).

8. Postoperative management

- Antibiotics
- Control of diabetes (if patient is diabetic)
- Regular dressings of the wound with antiseptic agents.

9. Postoperative complications

- During the process of breaking the loculi, vessels underneath may be injured causing haematoma which requires drainage. Otherwise no specific complications.
- Injury to vessels or nerves can occur if basic principles of drainage of an abscess are not followed. When an abscess is located over the major vessel, as in axilla or neck, **do not make a stab incision**. An incision is made on the skin and subcutaneous tissue and sinus forceps is introduced. Later, it is treated like the treatment of an abscess. This

method is followed to avoid injury to major vessels. It is also indicated in parotid abscess to avoid damage to facial nerve. This is called **Hilton's method of drainage**.

10. Advice at discharge

Control of diabetes (if present)

INCISION AND DRAINAGE OF BREAST ABSCESS

1. Indication

Breast abscess (Key Box 53.5)

2. Contraindication

None

3. Position of the patient

Supine

4. Anaesthesia

GA

5. Preparation

Iodine and spirit

6. Procedure (Figs 53.11 and 53.12)

A 5–6 cm long semicircular incision is made over the swelling where there is maximum tenderness. It is drained just like pyogenic abscess. Another stab incision is made in the dependent position and corrugated rubber drain/plastic is brought out through this incision.

7. Closure

- If infection is very severe, **do not close the incision**.
- Otherwise, main wound is sutured and corrugated red rubber drain is brought down at the dependent position. Once the drainage is minimal, the drain is removed.

PEARLS OF WISDOM

Minor breast abscess need not be drained. One or two aspirations may be curative in many cases.

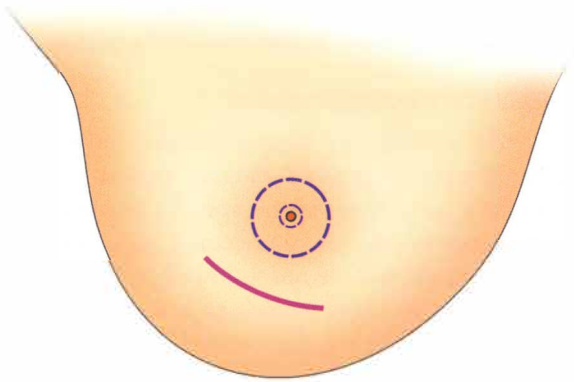


Fig. 53.11: Incision for breast abscess

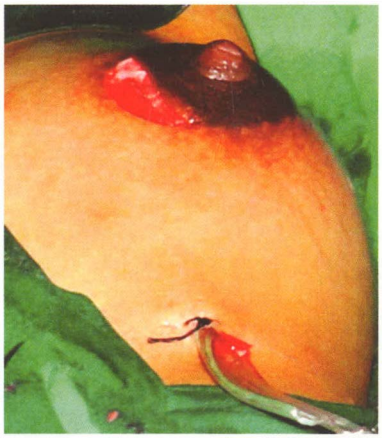


Fig. 53.12: Drainage and usage of corrugated rubber drain

KEY BOX 53.5

BREAST ABSCESS DRAINAGE

- GA is preferred
- Do not wait for fluctuation
- Throbbing pain is an indication for surgery
- Small curved incision
- Keep in mind, mastitis carcinomatosa

Ultrasound-guided aspiration of breast abscess should be done first especially in unilocular breast abscess.

8. Postoperative management

- NPO for about 6 hours
- Antibiotic of choice is **cloxacillin** 500 mg 6th hourly because the common organism is *Staphylococcus aureus*.
- It may take 7–15 days for complete healing.
- One should not wait for fluctuation to develop in a breast abscess. If pain and tenderness does not subside by 48 hours, breast abscess is incised. Otherwise, breast tissue gets damaged.

9. Postoperative complications

Haematoma needs evacuation

10. Advice at discharge

Lactating women should clean the nipple after every breast feed and keep it clean.

PEARLS OF WISDOM

Do not use radial incision in breast surgery.

CIRCUMCISION

Circumcision refers to removal of the preputial skin.

1. Indications

a. Ritual: Religious
b. Phimosis

2. Contraindication

Hypospadias

PEARLS OF WISDOM

Preputial skin is required for repair of hypospadias.

3. Position of the patient

- Supine

4. Anaesthesia

a. In children—GA
b. In adults—LA

5. Preparation of the parts

Savlon and spirit

PEARLS OF WISDOM

Use plain lignocaine (without adrenaline) for LA during circumcision. Dose: 2% lignocaine 10–15 ml.

6. Procedure

In adults (Fig. 53.13)

- Skin of the tip of the penis is held in two places by using artery forceps, prepuce is separated from the glans and is slit up in mid-dorsal line to a point a little beyond the middle of the glans.
- Preputial layers are trimmed away in a line parallel to the corona. ***On the ventral surface, frenular artery needs to be ligated by using figure of 8 stitch.*** Two layers of prepuce are united by interrupted fine chromic catgut sutures/vicryl. Dressings are applied.

In children (Fig. 53.14)

- Prepuce is held by two artery forceps and gentle traction is applied. A small artery clamp is applied distal to the glans and skin distal to the clamp is removed.
- Once clamp is removed, bleeding points are identified and ligated.
- Two layers of prepuce are approximated by using chromic catgut.

7. Closure

Two layers of prepuce by using chromic catgut/vicryl.

8. Postoperative management

- Sedatives and analgesics
- Antibiotics
- Removal of sutures is very painful. Hence, **do not use nonabsorbable sutures**.

9. Postoperative complications

a. Injury to the glans penis can occur when there are extensive adhesions between prepuce and glans. It needs suturing.

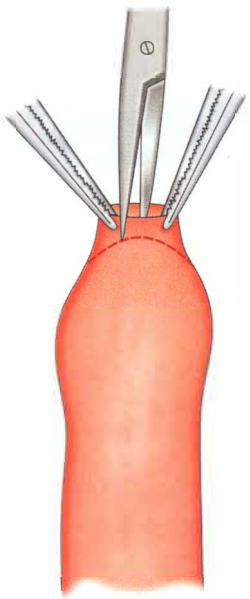


Fig. 53.13: Circumcision in adults

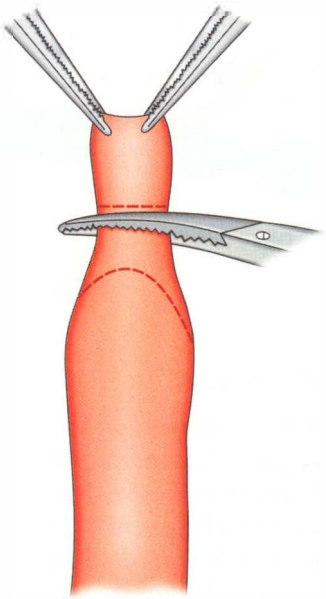


Fig. 53.14: Circumcision in children

b. **Haematoma:** Due to injury to the corpora cavernosa or due to the bleeding from cut edges.
c. **Tension at suture line** if too much skin is removed. This may cause painful erection at a later date.

10. Advice at discharge

This surgery in adults is done on an outpatient basis. Patients are discharged within a few hours. Hence, patients are advised to report if there is bleeding and also not to wet the area for 2–3 days.

VENESECTION OR CUT DOWN

1. Indications

- Shock: Hypovolaemic, haemorrhagic, burns, etc.
- When peripheral veins are not visible due to shock, burns or massive haemorrhage, an incision is made in the anatomical sites of the vein. The vein is identified, isolated and cannulated for transfusion of fluids. This procedure is called *venesection* or *cut down* (Fig. 53.15).

2. Contraindication

No

3. Position of the patient

Supine

4. Anaesthesia

Local infiltration by using 2% lignocaine 3–5 ml

5. Preparation

Iodine and spirit

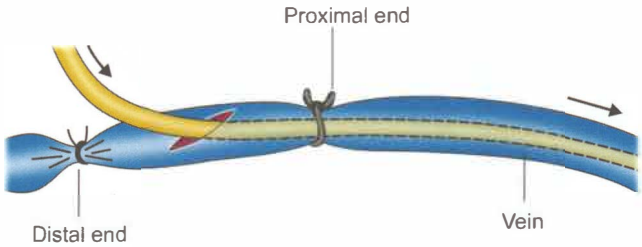


Fig. 53.15: Venesection

6. Procedure

Cephalic vein cut down is the most popular and an ideal procedure. A transverse incision about 5 cm is made in the deltopectoral groove. The cephalic vein is isolated and the distal end of the vein is ligated so that venous blood does not leak. A nick is made in the vein, through which a sufficient sized cannula (infant feeding tube can be used) is introduced. A silk ligature is applied above, just tight enough to hold cannula in place. Free flow of venous blood in the cannula indicates that it is inside the vein. The cannula is advanced further for about 10–15 cm. It is connected to IV line containing fluid (Fig. 53.15).

Precautions

1. **Neither air bubbles should be injected** nor should they be present in the drip set, to avoid air embolism.
2. **Upper ligature should not be tight.** It may obstruct the flow of fluids.
3. **Strict antiseptic principles** must be followed to avoid septicaemia.

Other veins selected for cut down

- Basilic vein in arm
- Cubital vein at the elbow
- Long saphenous vein in the leg. Veins in the leg, as far as possible, should be avoided to prevent deep vein thrombosis.

7. Closure

Skin—interrupted silk

8. Postoperative management

- Care of wound by dressing
- To avoid air bubbles in the drip set

9. Postoperative complications

- Infection, chills, rigors and septicaemia
- Air embolism

10. Advice at discharge

Nil

Advantages of cephalic vein cut down

1. Reliable vein and easy to do
2. If cannula is advanced into the right heart, CVP can be measured.
3. Mobility of the patient is not restricted.
4. Substances which cannot be given in a peripheral vein, such as 50% dextrose, lipids, amino acids, etc. can be given without risk of thrombosis of the vein, for hyperalimentation purposes.

PEARLS OF WISDOM

Cannulate vein, not an artery for venesection. Thin-walled nonpulsatile blue structure is vein.

VASECTOMY

Division and removal of a part of the vas deferens is vasectomy (Fig. 53.16).

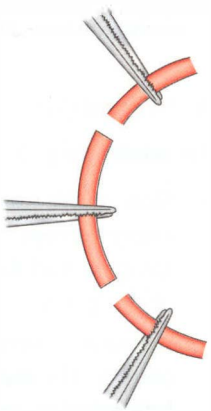


Fig. 53.16: Division of vas deferens by three clamp method

1. Indications

- Family planning
- To prevent epididymo-orchitis after prostatectomy. (Nowadays not routinely done)

2. Contraindications

- **Relative:** Tuberculosis epididymo-orchitis. The incision may result in a nonhealing sinus. Hence, control of tuberculosis is done first followed by vasectomy.
- **Absolute:** Suspicion of testicular malignancy.

3. Position of the patient

Supine

4. Anaesthesia

Local anaesthesia using 3–5 ml of 2% lignocaine.

5. Preparation of the parts

- Savlon and spirit
- Iodine is better avoided

6. Procedure

Feeling the vas deferens

After cleaning and draping, the vas is felt, at the root of scrotum between the index finger and thumb. It feels like a cord. Lignocaine is infiltrated and wait for 1–2 minutes for lignocaine to act.

Incision

An incision of 2–4 cm is made in root of scrotum and it is deepened through layers of scrotum. An 'Allis forceps' is introduced within the incision and spermatic cord is held.

During this step, fingers of the other hand help in guiding/locating/stabilising the cord. The coverings of the cord are incised.

Precautions

- Do not damage testicular vessels
- Vas is separated. It is confirmed by its white colour, and it feels like a cord.
- Division of vas by three clamp method (Fig. 53.16).
- Vas is cut in two places A and B so that a piece of vas is removed, which can be sent for histopathology to confirm that it is vas.
- Since a piece of vas is removed, reunion of the cut ends will not occur.
- The two cut ends of vas are doubly ligated by using silk.

7. Closure

The skin is closed by absorbable one or two sutures so that removal not required.

8. Postoperative management

- Rest for a few hours
- Antibiotics and analgesics

PEARLS OF WISDOM

The procedure is repeated on the other side.

9. Postoperative complications

- Injury to the vessels, resulting in a large haematoma.
- Infection
- Testicular atrophy can occur a few years later. It is due to immunological reaction rather than disuse atrophy.

10. Advice at discharge

To use other methods of family planning for two months while having sexual intercourse, as some sperms may be present in the distal end of the vas and seminal vesicle.

PEARLS OF WISDOM

Vasectomy being a part of family planning project, every student should be familiar with this.

No scalpel vasectomy

- It is a novel technique to do vasectomy through one single puncture which does not require any suturing. It is less traumatic than conventional vasectomy and shortens recovery time.
- The procedure is done with LA.
- A special instrument is used to puncture the scrotum and grasp the vas deferens. Vas is then cut and through the same puncture, the other side is also operated.

TRACHEOSTOMY

An opening made in the trachea is tracheostomy

1. Indications

a. Emergency

- Choking of the larynx due to dentures, foreign bodies fish bones, etc.
- Stridor due to diphtheria, carcinoma larynx and bilatera recurrent laryngeal nerve paralysis after thyroidectomy

b. Elective

- Coma
- Tetanus
- Barbiturate poisoning
- Head injuries
- Pulmonary insufficiency

2. Contraindications

Anaplastic carcinoma thyroid patients presenting with stridor due to infiltration of growth into trachea. It may not be possible to do a tracheostomy or an attempt to do tracheostomy may result in the growth fungating through the incision (which is best avoided). In such patients, **endotracheal intubation** is done if possible. If not possible, no other intervention is done.

3. Position of the patient

Supine with extension of the neck and head by keeping a sandbag or a pillow under the shoulders.

4. Anaesthesia

Local infiltration anaesthesia

5. Preparation of the parts

Iodine and spirit

6. Procedure

- **Incision:** Transverse curved incision for about 3–4 cm is made at the level of 2nd tracheal ring.
- **Dissection:** Skin, subcutaneous tissue and deep fascia are incised. Isthmus of thyroid is separated.
- **Procedure:** A transverse cut is made in the 2nd tracheal cartilage, its edge is held with Allis forceps and a small cuff of cartilage is removed. 'Cricoid hook' can be used to stabilise the trachea (found more useful in children).
- A suitable-sized tracheostomy tube is introduced within.
- The cuff of tracheostomy tube is inflated by using 2–5 ml of air and is held in place by passing a tape around the neck.
- Confirm that the **tube** is in the **trachea**, not in the subcutaneous plane.
- Confirm **air entry** on both sides of lung.

- Urinary bladder is closed with absorbable 2–0 chromic catgut sutures.
- A corrugated drainage tube is kept in the prevesical space.

7. Closure

- Split rectus muscles are approximated by using catgut sutures.
- Anterior rectus sheath/linea alba—by nonabsorbable sutures.
- Subcutaneous tissue—chromic catgut
- Skin—silk

8. Postoperative management

- Antibiotics
- Analgesics

9. Postoperative complications

a. Wound infection
b. Haemorrhage: Bladder wash is given till the contents are clear. Rarely, it may need re-exploration and control of the bleeding.
c. If urinary leakage occurs, it is usually minor.

10. Advice at discharge

Nil

PEARLS OF WISDOM

This operation has become less popular because of suprapubic catheterisation which is a simple procedure. This operation will help in case of large stones which cannot be removed or crushed by transurethral approach.

THYROIDECTOMY

1. Indications

All goitres with symptoms—MNG, toxic goitre, colloid goitre and malignant goitre.

2. Contraindications

Asymptomatic goitre, Hashimoto's thyroiditis, anaplastic carcinoma thyroid.

3. Position of the patient

- Supine with extended neck by keeping a sandbag under the shoulders.
- Head end of the patient is elevated to about 30° to reduce venous congestion. This position is called **anti-Trendelenburg position**.

4. Anaesthesia

GA.

5. Preparation of the parts

Iodine and spirit

6. Procedure

Incision (Fig. 53.19)

6–8 cm collar neck incision or crease incision

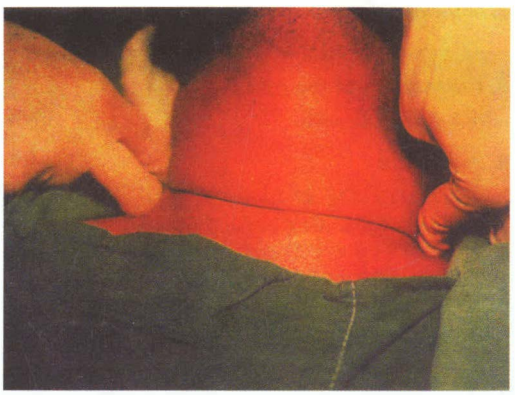


Fig. 53.19: Incision for thyroidectomy being marked with silk thread prior to incision

Layers opened

- Skin, platysma, subcutaneous tissue in the line of incision
- Deep fascia is incised vertically.
- Strap muscles are separated (can be cut in very large goitres).
- Pretracheal fascia is incised.
- Thyroid gland is mobilised by using blunt dissection.
- Assess the entire gland to know whether it is a solitary nodule or multinodular goitre.
- One of the lobes is mobilised by dividing middle thyroid vein (single, short, thin, vein).
- Then, upper pole is dissected. This pedicle contains superior thyroid artery and veins. They are ligated and divided in between. **Please apply double ligature proximally.**

PEARLS OF WISDOM

All major arteries should be ligated twice proximally. Example: Superior thyroid artery, facial artery, left gastric artery, cystic artery, renal artery.

- Upper pole should be ligated as close to the gland as possible to avoid damage to external laryngeal nerve.
- Inferior thyroid artery used to be ligated[1] well away from the gland. It has a horizontal course. It is thick and pulsatile. Then, the brances of inferior thyroid artery are ligated. This

[1]Today, branches of inferior thyroid artery are ligated—not the main artery so as to preserve blood supply to parathyroid gland.

will avoid injury to recurrent laryngeal nerve and it will prevent hypoparathyroidism also. Multiple veins, present in the lower pole are ligated and divided.

- Isthmus is separated from trachea, both above and below.
- In subtotal thyroidectomy the entire isthmus, parts of the right and left lobes are removed in flush with tracheal surface, leaving behind tissue in the tracheo-oesophageal groove to protect recurrent laryngeal nerve and parathyroid gland. Cut edges of thyroid gland are sutured by using vicryl sutures (Fig. 53.20). In total thyroidectomy almost entire gland is removed (Fig. 53.21).

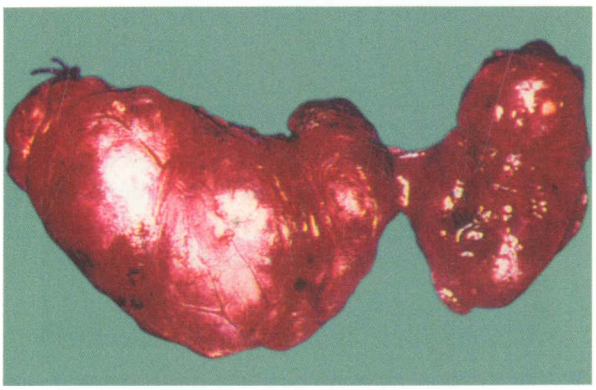


Fig. 53.20: Subtotal thyroidectomy for MNG

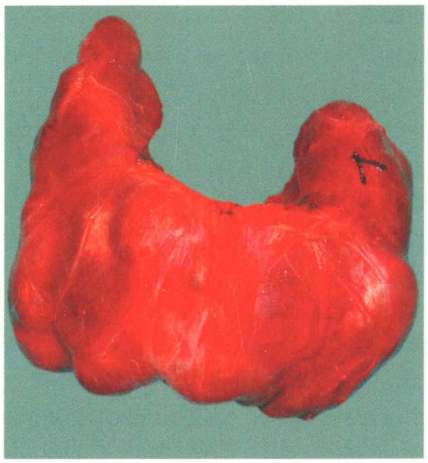


Fig. 53.21: Total thyroidectomy specimen for papillary carcinoma thyroid

Precautions

- Any structure directly entering the gland is unlikely to be RLN and hence, can safely be divided.
- Recurrent laryngeal nerve enters the thyrohyoid membrane, after running a vertical course, in the tracheo-oesophageal groove.

7. Closure

- A suction drain is kept in the thyroid bed.
- Deep fascia—continuous vicryl
- Subcutaneous fat—vicryl
- Skin—interrupted silk/subcuticular sutures
- A bandage is applied

8. Postoperative management

- NPO for 6–8 hours followed by liquid diet
- Antibiotics are not necessary
- **Head end must be elevated** to reduce oedema of the wound
- In toxic goitres, propranolol must be continued after surgery and slowly tapered over a week
- Blood transfusion depending upon blood loss
- Drain removal after 2–3 days (once it stops draining)
- Suture removal after 4–5 days

9. Postoperative complications

(for details *see* Chapter 19, page 310)

1. **Haemorrhage: Tension haematoma.** Reactionary haemorrhage is due to slipping of ligature due to coughing, hypertension, etc. If it is alarming, deep fascial sutures have to be opened, haematoma drained and haemostasis has to be achieved.
2. **Thyrotoxic crisis** in patients with toxic goitre
3. **Tracheomalacia**—resulting in stridor
4. Recurrent laryngeal nerve paralysis
5. Hypothyroidism
6. Hypoparathyroidism
7. Wound infection

Different types of thyroidectomy (Table 53.2)

10. Advice at discharge

This depends on the type of indication for thyroid surgery, e.g. those who undergo subtotal thyroidectomy for thyrotoxicosis have to be closely followed for recurrent thyrotoxicosis or hypothyroidism. If calcium levels are low, it has to be supplemented.

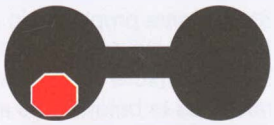
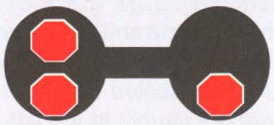
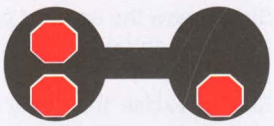
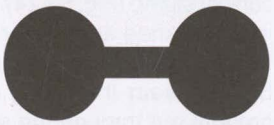
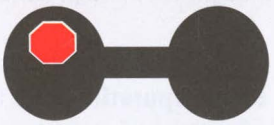
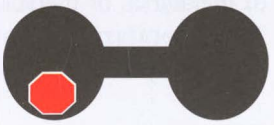
PEARLS OF WISDOM

Thyroidectomy is an operation which provides a surgeon to demonstrate his skills and 'meticulous' capacity.

What is Zuckerkandl's tubercle?

- **Zuckerkandl's tubercle** is a pyramidal extension of the thyroid gland, located at the most posterior side of each lobe.
- The structure is important in thyroid surgery as it is closely related to the recurrent laryngeal nerve, the inferior thyroid artery, Berry's ligament and the parathyroid glands.
- It is also important to remove this *in toto* while doing thyroidectomy for malignancies.

Table 53. 2 Different types of thyroidectomy

Diseases	Before surgery	After surgery	Name of the operation
1. Solitary nodule (benign)			Hemithyroidectomy means removal of one lobe with isthmus (*see* page 308)
2. Multinodular goitre			Subtotal thyroidectomy (*see* page 287)
3. Toxic multinodular goitre			Subtotal thyroidectomy (*see* page 292)
4. Primary thyrotoxicosis			Subtotal thyroidectomy (*see* page 292)
5. Malignant neoplasm			Near total thyroidectomy (*see* page 297)
6. Malignant neoplasm—high-grade carcinoma			Total thyroidectomy (*see* page 297)

AMPUTATIONS

Definitions/terminologies of amputation

- End bearing: Weight is taken by the body.
- Non-end bearing: Here weight is taken by the joint.
- *Guillotine* amputation: Here no flaps are raised, all the tissues are divided at the same level and the stump is kept open.
- Formal amputation: In this case depending upon the indications and the decisions taken by the surgeon, amputation is done with closure of the stump.

Indications

1. Vitality of the part is destroyed by injury or disease—***dead limb***
2. Life of patient is threatened by spread of a local condition—***deadly limb.***

   Examples: Gas gangrene, extensive melanoma
3. Patient may be better served by an artificial limb because of deformity or paralysis—***deformed limb. In such cases better to amputate and fit in an artificial limb.***
4. Dying limb—acutely ischaemic limb, late presentation.

Optimum levels of amputation

Level of amputation depends not only upon the extent of disease but also function desired in the remaining stump. This differs markedly in the upper and lower limbs.

Ideal stump

- Should have ideal length for proper fitting of prosthesis. Examples: Below knee: 8 to 12 cm from tibial tuberosity, above knee: 23 cm from greater trochanter and above and below elbow 20 cm stump.
- Should be conical and rounded
- Should not be tender
- Should have adequate muscle padding so that its movements are adequate.
- Should have adequate blood supply so that it heals with primary intention in the postoperative period.
- Should have a thin scar which should not interfere with prosthetic function.
- Should not have any redundant soft tissue hanging.
- Skin and scar should not be adhered to the underlying tissue.

Incisions : Depending upon the site of the level of amputation and keeping in mind the blood supply of the part, different types of incision are given. They are as follows:

- *Racquet incision:* This is used in amputation for digits or toes.
- Elliptical or oval incision is given for metatarsal amputations.
- Circular incision is given especially in *Guillotine* amputation.
- U shaped incisions: These are given to raise flaps—anterior and posterior flaps as in below knee or above knee. By convention equal flaps are used for above knee and a long posterior and short anterior flaps are used for below knee amputation. This is because, vascularity of the posterior flap is good below the knee due to bulky muscles with good blood supply when compared to the thin, muscle less anterior flap (*see* ten commandments).

AMPUTATIONS IN LEG

Skeleton of foot: To have a better understanding of amputations kindly study Fig. 53.22 and 53.23 first.

- One of the common indications for lower limb amputations is diabetic ulcer/ gangrene foot. Various types and various levels of amputations are done with the main aim is to conserve as much as possible. However, when the limb is a useless limb, a below knee or an above knee amputation is done depending upon the seriousness of the problem.

TEN COMMANDMENTS: GENERAL PRINCIPLES IN AMPUTATIONS

1. Should mark the incision
2. Should give prophylactic antibiotics
3. Should avoid **tourniquet** in arterial occlusive diseases.
4. Should ensure adequate blood supply to the flaps—if raised as in below knee and above knee amputations.
5. Should ligate the blood vessels securely to avoid haematoma and then infection in the postoperative period.
6. Should not clamp the nerves but they are pulled down and transected as high as possible so that nerve ends are not caught in the suture line.
7. Should saw the anterior part of the bone obliquely to give a smooth anterior bevel which prevents pressure necrosis of the flap.
8. Should excise the bulky muscles so as to give a good conical stump (Fig. 53.24). Example: Excise soleus muscle in below knee amputations.
9. Should use absorbable sutures to unite the muscle ends.
10. Should drain the cavity with a suction drain which is brought out through the skin clear of the wound.

1. **Ray amputation:** It is amputation of the toe with head of metatarsal or metacarpals.
2. **Transmetatarsal/metacarpal amputation:** It is called Gilles' amputation. When multiple toes are involved with

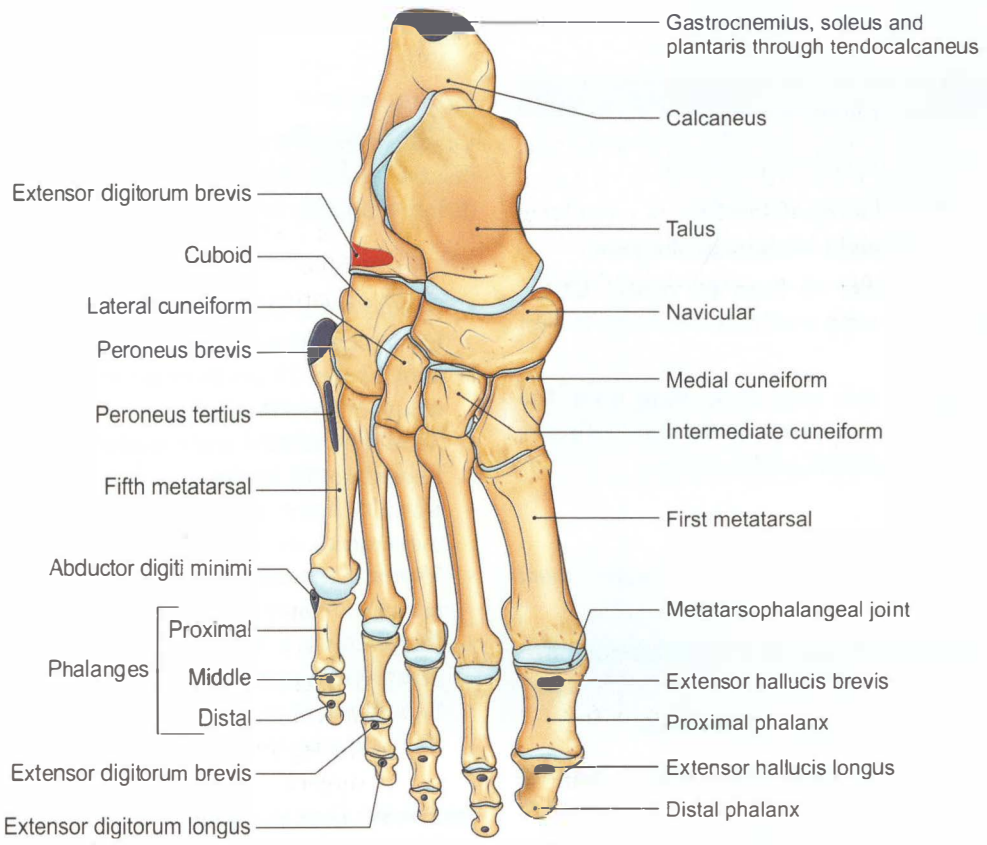


Fig. 53.22: Skeleton of the foot as seen from the dorsal aspect

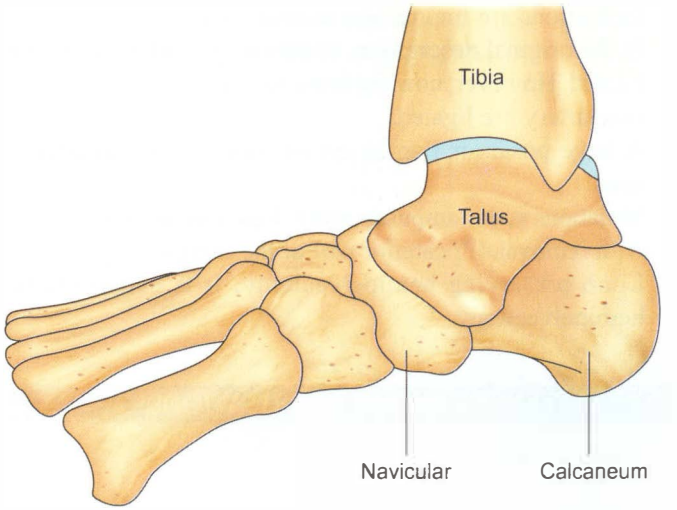


Fig. 53.23: Tarsal bones

gangrene as in vasculitis syndromes or in diabetic patients, amputation is done through metatarsal bones—proximal to the neck, distal to the base. Long volar flap is created and sutured to the dorsal skin.

3. **Lisfranc's amputation (tarsometatarsal amputation):** Tarsometatarsal articulations are called Lisfranc joint. The bones forming these are the first, second, and third cuneiforms, and the cuboid, which articulate with the bases of the metatarsal bones. The bones are connected by dorsal, plantar, and interosseous ligaments. These ligaments have to be divided. A long volar flap is used. Patient needs a surgical boot.
4. **Chopart's amputation:** Francis Chopart first described disarticulation through midtarsal joint. It is midtarsal amputation. Disarticulation of the foot is completed through talonavicular joint and through calcaneocuboid joint. Thus, Chopart amputation removes the forefoot and midfoot, saving talus and calcaneus. Tibialis anterior muscle is sutured to the drilled talus bone.
   - **Contraindication:** Ischaemic feet as in atherosclerosis.
   - **Disadvantages:** It is a very unstable amputation, because most of the tendons supporting the foot will be removed. Thus, it will go for equinus and must usually be fitted with a prosthesis that extends up to the patellar tendon level.
5. **Syme's amputation:**
   - The tibia and fibula are divided at or immediately above the level of ankle joint and their ends are covered with a single flap obtained from heel.
   - The end of the stump is at a height of about 6–8 cm from the ground.
   - 50% of people will be able to walk on the stump without prosthesis.
   - It is of value in patients who do not have access to modern artificial limbs.

Fig. 53.24: Conical stump

   - **Pirgroff's modification of Syme's amputation** retain a small portion of calcaneum in the flap obtained fron heel (Fig. 53.24).
   - Heel flap is supplied by medial and lateral calcanea vessels, both are branches of posterior tibial artery.
   - Those who will not be able to walk after thi amputation, can be fitted with elephant boot.
6. **Below knee amputation:**
   - It is the operation of choice when it is not possible t preserve the foot or heel.
   - The ideal length of the tibial stump is 14 cm.
   - Minimum length required to fit an artificial leg is 8 cm Stump shorter than this tends to slip out of the socke of an artificial limb.
   - The stump is covered by creating long posterior flap.
   - This is the amputation commonly done in patients wh are in severe sepsis involving the leg with uncontrolled diabetes and life is in danger.
   - All the rules mentioned above in ten commandments are followed here such as division of the nerve, flap vascularity, reduction of bulky muscles and the anterior scar, thus prosthesis will not cause discomfort while walking.
   - Advantages of below amputation include greater range of movements without limp and without support.
   - This amputation is also called Burgess amputation.
   - POP cast should be put to present contractures (Fig. 53.25).
7. **Amputations through thigh**
   - Ideal length is 25–30 cm as measured from tip of trochanter.
   - It is done when it is not possible to save at least 8 cm of tibia as in some cases of diabetes or spreading infections of the leg and when muscles involved are not bleeding at surgery.
   - When this amputation is done in children, as much length as possible should be preserved (growing epiphysis of femur is at lower end).

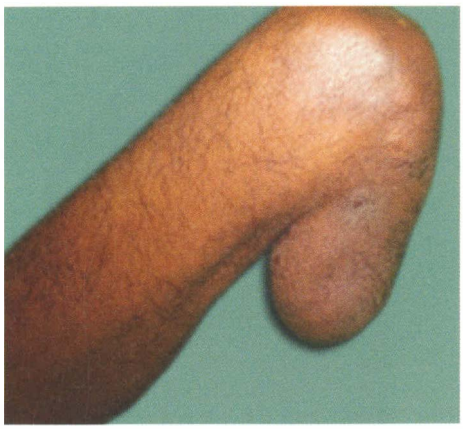


Fig. 53.25: Amputation contracture

- Unlike, below knee amputation, equal flaps are raised—anterior and posterior.
- Any length less than 10 cm of femur will not help. In such cases, hip disarticulation is done.
- In peripheral arterial occlusive disease, an attempt is made first by raising below knee flaps. If edges of the skin flaps do not bleed, better to go ahead with above knee flaps because vascularity of above knee flaps are better. Above knee amputation stump healing is better than below knee.
- Disadvantages of this amputation are difficult rehabilitation (not easy), prosthesis fitting is not good, invariably patient needs one more support (Fig. 53.26).

8. Hip disarticulation

- When it is not possible to get minimum of 10 cm length of stump of the femur, hip disarticulation is done. This situation can occur in trauma or malignancies to get a wide clearance. Examples: Sarcomas or in cases of malignant melanomas.
- Usually a single posterior flap is raised—Solcum's approach.
- Anterior approach can also be used (2nd option)—Boyd's approach

9. Hindquarter amputation

- In this amputation—one side of pelvis with innominate bone, pubis, muscles and vessels are removed. Hence, it is called hemipelvectomy today.

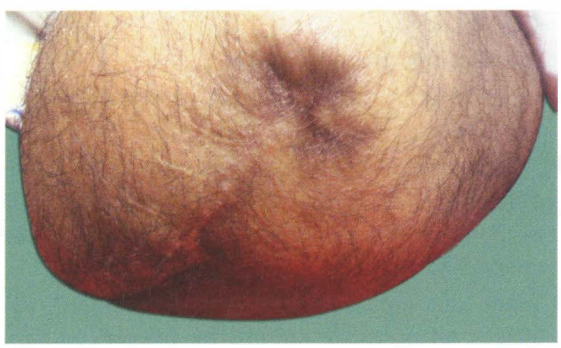


Fig. 53.26: Infected scar

- Indications are trauma and tumour (malignancy).
- In the original description, common iliac artery used to be ligated. However, now the branches of external and internal iliac artery are ligated.
- A large posterior flap based on superior gluteal artery is used.
- Variations in this amputation are: Extended hemipelvectomy with removal of posterior part of the sacrum.
- Limb preserving hemipelvectomy: It is called internal hemipelvectomy.

UPPER LIMB AMPUTATIONS

GENERAL PRINCIPLES

- Conserve as much tissue as possible.
- Skin closure should not be under tension.
- Soft tissue cover over bony stump is desirable. Otherwise, painful adherent scar will result.
- Amputation through middle or terminal phalanx is preferred to disarticulation at interphalangeal joints since attachment of flexor tendons is thereby preserved.
- Every effort should be made to preserve as much of the thumb.

AMPUTATION THROUGH FOREARM AND UPPER ARM

- Ideal stump is 16–20 cm measured from olecranon.
- Stump less than 8 cm is useless for transmitting movement to an artificial elbow joint.
- A stump measuring 20 cm from acromion is ideal for fitting prosthesis.

Krukenberg's amputation: In this amputation, a gap is created between radius and ulna like a claw. It helps in holding objects.

Interscapulothoracic amputation (forequarter amputation)

- Indications are for malignancy involving axial skeleton such as sarcoma. Sepsis involving the upper limb is another indication such as gas gangrene.
- It is a very radical mutilating operation, hence all possible limb saving attempts should be done first.
- Entire upper limb with scapula and lateral 2/3rds of the clavicle with all the muscles attached to it are removed.

Complications following amputation

1. **Wound infection:** Especially it is common in amputations done for diabetic gangrene cases. Stitches may have to be opened to release pus followed by secondary suturing at a later date.
2. **Flap necrosis:** It is a common complication because of several reasons important one being decreased blood supply

to the limb either due to arterial occlusive disease or due to diabetes. Necrotic skin and subcutaneous tissues should be removed followed by secondary suturing at a later date. Hence, blood supply of the flap has to be kept in mind when raising the flaps.

3. Stump ulcers are common in the initial stages of wearing artificial limbs.
4. **Contracture:** If the artificial limb is not fitted, the stump will develop flexion contracture.
5. **Amputation neuroma:** This is an end neuroma. The cut end of the nerve is entrapped in the scar tissue and gives rise to pain. To avoid this, nerve end is pulled and cut so that after division of the nerve, the end gets retracted.
6. **Phantom limb:** A **phantom limb** is the sensation that a missing limb is attached to the body. Approximately 60 to 80% of individuals with an amputation experience phantom sensations in their amputated limb, and the majority of the sensations are painful. It is probably due to presence of a severe pain at the amputated site before surgery and the corresponding site in the brain has registered this sensation.

OPEN CHOLECYSTECTOMY

In the vast majority of the cases gall bladder is removed by laparoscopic route. Details have been given in the gall bladder chapter. In this chapter we will be studying gall bladder removed through open method—after doing a laparotomy.

Defintion

Removal of the diseased gall bladder by a laparotomy.

1. Indications

Laparoscopic cholecystectomy is now the gold standard for cholecystectomy. However, the role of open cholecystectomy is present when laparoscopic cholecystectomy fails due to extensive adhesions, **excessive bleeding**, CBD injury, impacted gall bladder, etc. (complications of laparoscopic cholecystectomy)

- Symptomatic gall stones
- Acute/chronic/acalculous cholecystitis
- Empyema gall bladder
- Mucocoele of gall bladder
- Asymptomatic gall stones—patients with high risk such as diabetes, haemolytic anaemias such as sickle cell anaemias and hereditary spherocytosis.

2. Contraindications

- Unfit for surgery
- Chronically debilitated patients

3. Position of the patient

Supine. In laparoscopic cholecystectomy, the head end is elevated and a slight tilt is given to left side so that omentum and bowel fall away from the operating field.

4. Anaesthesia

GA

5. Preparation of the parts

From level of nipple to lower abdomen, parts are cleaned wit povidone-iodine and spirit.

6. Surgical procedure

- **Incision:** Right subcostal incision (**Kocher's incision** preferred/right paramedian.
- **Layers opened:** Skin, subcutaneous tissue, muscle (external oblique, internal oblique and transversu abdominis), preperitoneal fat and peritoneum.
- **Dissection**
  - After opening the abdomen, colon and stomach ar retracted away.
  - Fundus of the gall bladder is held with a sponge holdin forceps and retracted.
  - Assistant retracts the liver using a Deaver retractor.
  - **Calot's triangle is identified.** The cystic artery i identified, doubly ligated with 2.0 silk sutures and cut.
  - Cystic duct is now identified, skeletonised, doubly ligate with silk or vicryl sutures and cut.
  - The gall bladder is dissected off the gall bladder foss using electrocautery and haemostasis is achieved.
  - Rarely, fundus first approach: When Calot's triangl anatomy is not clear due to inflammation and adhesions the dissection is started from the fundus and proceede towards the cystic duct which is ligated in the end.
  - Intra-abdominal drain is placed.

7. Closure

- Inner muscle layer—No.1 prolene continuous interlocking.
- Outer muscle (2nd layer of muscles)—same suture.
- Subcutaneous layer—2.0 vicryl interrupted.
- Skin—2.0 ethilon/silk vertical mattress.

8. Postoperative management

- Nil per oral till patient passes flatus
- To continue antibiotics in diabetic patients
- Watch for hypotension (bleeding), tachycardia, abdominal distension, pain (bile leak).
- If drain is kept, it is usually removed within 2–3 days.
- To avoid having oily foods

9. Postoperative complications

- Infections and subphrenic abscess
- Bleeding from cystic artery
- Injury to CBD or hepatic duct—presents with jaundice in the postoperative period.
- Bile leak and fistulae
- Biliary stricture formations (late)
- Injuries to colon, duodenum and mesentery.

PEARLS OF WISDOM

If bile leak continues or CBD clipping is suspected, ask for ERCP and treat accordingly. Cystic duct stump leak is best treated by ERCP and stenting of CBD as early as possible.

10. Advice at discharge

- Not to strain for 30 days—to prevent incisional hernia developing later.
- To avoid fatty food
- To report if jaundice develops (CBD injury or retained stone in CBD) or fever which may be due to subphrenic collection. (It can be treated with ultrasound-guided aspiration.)

PEARLS OF WISDOM

Identification of Y junction at open surgery and identification of cystic duct joining the infundibulum at laparoscopic surgery (dilated portion resembling elephant trunk) are the key points, which will help in avoiding bile duct injuries to a very large extent.

VAGOTOMY GASTROJEJUNOSTOMY (GJ)

Vagotomy GJ, as it was called, is a procedure that was commonly performed by surgeons until the invention of proton pump inhibitors increasing awareness and advent of upper GI endoscopies. Earlier, the incidence and complications of peptic ulcer were high and the commonly performed surgery for peptic ulcer was vagotomy GJ. It was also done for the complications of gastric ulcers such as gastric outlet obstruction due to strictures. However, nowaday this procedure is rarely done.

1. Indications

- Symptomatic peptic ulcer disease not responding to medical management
- Complications of gastric ulcers such as stenosis and bleeding.

2. Contraindication

Vagotomy needs a bit of dissection near the hiatus. Hence a risk of mediastinitis is present if vagotomy is done in cases of perforation. Hence a simple closure of perforation is done in emergency situations.

3. Position of the patient

Supine

4. Anaesthesia

G/A

5. Preparation of the parts

From level of nipple to lower abdomen, parts are cleaned with povidone iodine and spirit.

6. Surgical procedure

- **Incision**—upper midline
- **Layers opened**—skin, subcutaneous tissue, linea alba, preperitoneal fat, peritoneum.
- **Dissection**
  - After opening the abdomen, the pathology in the stomach or duodenum is noted and confirmed.
  - Gentle traction is given at the anterior stomach wall. The stomach is delivered out of the wound.
  - The oesophagus is palpated with the *in situ* nasogastric tube between the thumb and the fingers.
  - The peritoneum over the overlying distal oesophagus is incised and the oesophagus is gently mobilised. The oesophagus is encircled with a Penrose drain and lifted to visualise the anterior vagus. Once identified, it is cut after ligating or applying clips. A 2 cm portion of the nerve may be excised.
  - Similarly, the posterior nerve is found as a taut band between the right crus of diaphragm and the oesophagus which is identified and cut.
  - The duodenogastric junction is identified after lifting the transverse colon and its mesentery.
  - The first loop of the jejunum (1 foot from the DJ) is taken and gastrojejunal anastomosis is performed in 2 layers—inner full thickness continuous suture with 3.0 vicyrl and outer seromuscular interrupted sutures with 3–0 silk. The loop is usually taken posterior to the transverse colon through a surgically made rent in the transverse mesocolon (retrocolic) and is isoperistaltic.

7. Closure

- Peritoneum along with linea alba is sutured with no 1 prolene or loop ethilon continuous interlocking suture.
- Subcutaneous —2.0 vicryl interrupted sutures.
- Skin —2.0 ethilon vertical mattress sutures.

8. Postoperative management

- NPO for 2 days till patient passes flatus—indication that no anastomotic leak. Ryle's tube is removed, then followed by clear fluids by mouth for 2–3 days followed by soft diet.
- Suture removal by 7–10 days
- Fluid and electrolytes have to be checked in the post-operative period.

9. Postoperative complications

- Postvagotomy diarrhoea, due to denervation of the gut.
- Afferent loop obstruction/stomal oedema.
- Gall stone formation due to denervation of gall bladder.
- Stomal ulcers due to bile reflux
- Bile reflux gastritis

10. Advice at discharge

- To avoid large heavy meals
- Small frequent feeds better

- Avoid spicy and oily foods

All details about complications of vagotomy and GJ are given in page 503.

EXCISION OF SWELLINGS

A. LIPOMA

1. Indications

- Large size (cosmesis/patient's wish)
- Recent rapid increase in size (sarcomatous change)
- Symptomatic naevo/neurolipomas
- Causing pressure symptoms based on site.

2. Contraindications

Strictly speaking, there are no contraindications. However, asymptomatic lipomas in a difficult location need not be excised. However, small the surgery may be, safety is important principle to be kept in mind.

3. Position of the patient

Supine/lateral/prone depending upon the location.

4. Anaesthesia

If small, under LA, if large regional anaesthesia or GA.

5. Preparation of the parts

Povidone-iodine and spirit

6. Surgical procedure

- **Incision:** A linear incision over the summit of the swelling is placed and flaps raised on both sides of the incision.
- **Layers opened:** Skin and some part of the subcutaneous tissue till the capsule of the swelling is encountered.
- **Dissection:** Using an artery forceps or a mosquito forceps (if a small swelling), a plane is created between the raised flaps and the capsule of the swelling. Pressure is given at the base of the swelling to deliver out the lipoma. A small vessel may be encountered as the base is being dissected that should be identified and cauterised or ligated. The specimen should be sent for histopathological evaluation.

7. Closure

If a large cavity is created due to excision of swelling, the excised skin flaps can be refreshed and excess skin can be removed. A few interrupted vicryl sutures can be placed to close subcutaneous layer. The skin is closed with 2.0 ethilon vertical mattress suture. Sometimes, a drain may have to be kept in the cavity.

8. Postoperative management

Rest to the part to prevent bleeding.

9. Postoperative complications

- Infection, bleeding
- Injury to vital structures around
- Seroma formation if large cavity remains

10. Advice at discharge

Suture removal after 7–10 days

B. SEBACEOUS CYST

1. Indications

- Infection
- Complications such as **Cock's peculiar tumour**, horn ar calcification.
- Patient's wish (cosmesis)

2. Contraindication

No specific contraindication

3. Position of the patient

Supine/lateral/prone depending upon the location.

4. Anaesthesia

Mostly LA. Multiple cysts over scalp and scrotum may requi GA or regional anaesthesia.

5. Preparation of the parts

Povidone-iodine and spirit.

6. Surgical procedure

- **Incision: Elliptical incision** around the summit of th swelling encircling the punctum.
- **Layers opened**
  - Incision should be superficial. Care should be taken nc to cut open the cyst wall.
  - The principle is to **completely excise the cyst** with it wall and the overlying punctum and a bit of th surrounding skin around the punctum.
- **Dissection**
  - A plane is created between the skin and the cyst wall carefully, preventing opening of the cyst wall.
  - An Allis forceps may be applied to the punctum and th elliptical skin to get a traction. Flaps need to be raise gradually on either sides of the incision and then delive the cyst *in toto*.
  - It the cyst wall opens up, the sebum is remove completely and an effort to remove all the cyst wall, i piecemeal, is made.

7. Closure

Single layer closure of the skin.

8. Postoperative management

To keep the wound clean—proper hygiene.

9. Postoperative complications

- Flap necrosis if too large a swelling and thin skin flaps
- Infection
- Recurrence, if cyst wall is not completely removed.

10. Advice at discharge

Suture removal after 7–10 days.

C. NEUROFIBROMA

1. Indications

- Cosmesis
- Symptoms of pain on pressure
- Pressure effects causing neurological deficits
- Sarcomatous changes

2. Contraindication

In von Recklinghausen's disease, only symptomatic neurofibromas should be removed.

3. Position of the patient

Supine/lateral/prone depending upon the location.

4. Anaesthesia

Mostly LA, sometimes GA

5. Preparation of the parts

Povidone-iodine and spirit.

6. Surgical procedure

- **Incision:** A linear incision over the summit of the swelling is placed and flaps raised on both sides of the incision.
- **Layers opened:** Skin and some part of the subcutaneous tissue till the capsule of the swelling is encountered.
- **Dissection:** Using an artery forceps or a mosquito forceps (if a small swelling), a plane is created between the raised flaps and the capsule of the swelling. Pressure is given at the base of the swelling to deliver out the neurofibroma. Care should be taken not to injure the underlying nerve while dissecting.

7. Closure

It can be closed in two layers, subcutaneous—vicryl, interrupted and skin 2.0 ethilon vertical mattress.

8. Postoperative management

Rest to the affected part, to keep the wound clean.

9. Postoperative complications

- Infection
- Injury to the nerve causing weakness, loss of sensations of the affected part.
- If partially left behind, recurrence and chance of sarcomatous changes.

10. Advice at discharge

Suture removal after 7–10 days.

Please note: In the following pages, a few more operations have been discussed which are generally not asked for MBBS students. However, knowledge of these may help you to secure more marks.

COLONIC SURGERY

RIGHT HEMICOLECTOMY

Indications

1. Carcinoma of the caecum or ascending colon
2. Tumours of appendix
3. Extended right hemicolectomy for carcinoma of hepatic flexure and proximal 1/3 transverse colon and for closed loop obstruction in carcinoma of the transverse colon.
4. Modified right hemicolectomy for tuberculosis, Crohn's disease involving terminal ileum (Fig. 53.27).

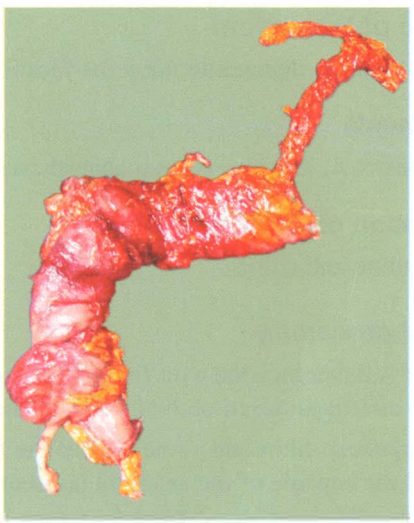


Fig. 53.27: Right hemicolectomy

Structures removed

1. Terminal 5–10 cm of ileum
2. Caecum
3. Ascending colon
4. Proximal 1/3rd of transverse colon with hepatic flexure.
5. Extended right hemicolectomy—all the above structures with the proximal two-thirds of the transverse colon.
6. **Modified right hemicolectomy**—all the above structures with variable length of hepatic flexure or proximal transverse colon preserved.

Vessels ligated

1. Ileocolic vessels
2. Right colic vessels

3. Right branch of middle colic vessels
4. Ileal vessels are ligated last

Surgical techniques

1. **Laparotomy** through midline/right paramedian incision.
2. **Exploration** of the abdomen for liver deposits, peritoneal deposits, ascites and other synchronous lesions.
3. **Assessment** of the tumour for site, extent, mobility, serosal involvement and local extension.
4. **Mobilisation of the right colon** done by incising along the avascular lateral peritoneal fold or **white line of Toldt** and rotating the caecum and the ascending colon anteriorly and medially.
5. **Retroperitoneum is entered** through the incision and dissection carried up towards the third and fourth part of duodenum.
6. Retroperitoneal structures encountered during dissection are the right kidney with **ureter, right gonadal vessels and duodenum**. Care must be taken to avoid injury to these structures.
7. **Turnbull's technique or no touch isolation technique**—early ligation of the vessels before manipulation of the tumour should be followed to prevent dissemination of the tumour cells during handling.
8. Ileocolic, right colic, right branch of middle colic and lastly ileal **vessels are isolated, ligated and divided.** Ileocolic and right colic vessels are ligated at the origin to include all the associated lymph nodes.
9. Ileum is transected at 5–10 cm from the ileocaecal junction and transverse colon at the junction of the proximal 1/3rd and distal 2/3rds. This is followed by an **ileotransverse colon anastomosis**.
10. Abdominal wall is closed in layers.

LEFT HEMICOLECTOMY

Indications

1. Carcinoma of the descending colon
2. Carcinoma of the splenic flexure
3. High-risk polyps

Structures removed

Carcinoma of the descending colon
1. Distal 1/3rd of transverse colon
2. Splenic flexure
3. Descending colon
4. Sigmoid colon

Carcinoma of the splenic flexure

1. Distal 2/3rds of transverse colon
2. Splenic flexure and descending colon

Vessels ligated

1. Left branch of middle colic vessels
2. Left colic vessels
3. Inferior mesenteric and sigmoidal vessels in case carcinoma of descending colon.

Surgical techniques

1. Laparotomy through midline/left paramedian incision.
2. Exploration of the abdomen for liver deposits, periton deposits, ascites and other synchronous lesions.
3. Assessment of the tumour for site, extent, mobility, sero involvement and local extension.
4. Mobilisation of the left colon done by incising along t avascular lateral peritoneal fold or white line of Toldt a rotating the descending colon and the sigmoid colon an riorly and medially. Splenic flexure is mobilised by dividi the gastrocolic ligaments and phrenicocolic ligaments.
5. **Retroperitoneum is entered** through the incision a dissection carried medially towards the ligament of Trie
6. Retroperitoneal structures encountered during dissecti are the **left kidney with ureter and left gonadal vesse** Care must be taken to avoid injury to these structures.
7. **Turnbull's technique** or no touch isolation technique early ligation of the vessels before manipulation of t tumour should be followed to prevent dissemination of t tumour cells during handling.
8. **Left colic and left branch of middle colic vessels** a isolated, ligated and divided in case of carcinoma of spler flexure. Inferior mesenteric and sigmoidal vessels are al ligated in case of carcinoma of descending colon.
9. **Level of colonic transection and anastomosis:**
   - Carcinoma of the splenic flexure: Proximally at t junction of right 1/3rd and left 2/3rds of transverse col and distally at the junction descending and sigmo colon. Colocolic anastomosis is done.
   - Carcinoma of the descending colon: Proximally at t junction of right 2/3rds and left 1/3rd of transverse col and distally at the rectosigmoid junction.
   - Colorectal anastomosis is done
10. **Abdominal wall is closed in layers**

TRANSVERSE COLECTOMY

Indication

Carcinoma of transverse colon

Structures removed

Whole of the transverse colon including the hepatic an splenic flexures.

Vessels ligated

1. Middle colic vessels
2. Left branch of right colic vessels
3. Right branch of left colic vessels

Surgical techniques

1. Laparotomy through midline incision
2. Exploration of the abdomen for liver deposits, peritoneal deposits, ascites and other synchronous lesions.
3. Assessment of the tumour for site, extent, mobility, serosal involvement and local extension.
4. Mobilisation of the hepatic flexure is done by incising along the right avascular lateral peritoneal fold or white line of Toldt and rotating the hepatic flexure anteriorly and downwards. Splenic flexure is mobilised by dividing the gastrocolic ligaments and phrenicocolic ligaments.
5. **Turnbull's technique or no touch isolation technique**—early ligation of the vessels before manipulation of the tumour should be followed to prevent dissemination of the tumour cells during handling.
6. Isolation of middle colic, left branch of right colic and right branch of left colic vessels done and ligated.
7. Proximal transection is done just proximal to the hepatic flexure and the distal transection is done just distal to the splenic flexure.
   - Colocolic anastomosis is done
8. Abdominal wall is closed in layers

SIGMOID COLECTOMY

Indications

1. Carcinoma sigmoid colon
2. Diverticular disease
3. Sigmoid volvulus

Structures removed

1. Sigmoid colon
2. Associated mesosigmoid
3. Lymph nodes in malignancy

Vessels ligated

1. Inferior mesenteric vessels distal to the origin of left colic vessels.
2. Sigmoidal vessels
3. Left branch of left colic vessels

Surgical techniques

1. **Laparotomy through midline**/left paramedian incision.
2. Exploration of the abdomen for **liver deposits**, peritoneal deposits, ascites and other synchronous lesions.
3. **Assessment of the tumour** for site, extent, mobility, serosal involvement and local extension.
4. **Mobilisation of the left colon** is done by incising along the avascular lateral peritoneal fold or white line of Toldt and rotating the descending colon and the sigmoid colon anteriorly and medially.
5. **Retroperitoneum is entered** through the incision and dissection carried medially towards the origin of inferior mesenteric artery.
6. Retroperitoneal structures encountered during dissection are the **left ureter and left gonadal vessels**. Care must be taken to avoid injury to these structures.
7. **Turnbull's technique or no touch isolation technique**—early ligation of the vessels before manipulation of the tumour should be followed to prevent dissemination of the tumour cells during handling.
8. **Inferior mesenteric vessels** distal to the origin of left colic vessels, sigmoidal vessels and left branch of left colic vessels done are isolated, ligated and divided.
9. **Colonic transection** done proximally at the junction of descending colon and sigmoid colon and distally at the rectosigmoid junction. Colorectal anastomosis is done.
10. Abdominal wall is **closed in layers**

LAPAROSCOPIC SURGERY

Introduction and history

- Laparoscopy made marked advances in the 1990s. Although the term minimally invasive surgery (MIS) is relatively recent, the history of its component parts is nearly 100 years old. What is considered the newest and most popular variety of MIS, laparoscopy, is, in fact the oldest.
- Primitive laparoscopy, placing a cystoscope within an inflated abdomen, was first performed by Kelling in 1901.
- In the late 1950s, Hopkins described the rod lens, a method of transmitting light through a solid quartz rod with no heat and a little light loss.
- Muhe in Germany began performing laparoscopic-assisted cholecystectomies in 1985.
- In 1987, Mouret and Dubious performed the first video-laparoscopy in France.
- The explosion of **video-assisted surgery** in the past 20 years was a result of the development of compact, high-resolution, charge-coupled devices (CCDs) that could be mounted on the internal end of flexible endoscopes or on the external end of a Hopkins telescope.
- Coupled with bright light sources, fibreoptic cables, and **high-resolution video monitors**, the video endoscope has changed our understanding of surgical anatomy and reshaped surgical practice.

Basic instrumentation

- **0° or 30° angled laparoscope** either 5 or 10 mm in diameter attached to camera connected to video source and monitor, ports for gas connection.
- **5-mm laparoscopic instruments** including Maryland dissector, blunt-tip dissecting forceps, cup-biopsy forceps, atraumatic grasping forceps, liver retractor, Babcocks forceps and scissors.
- **5 or 10 mm suction/irrigation device**
- **Laparoscopic ultrasound probe** (optional)

Equipment

1. **Telescope:** 30°, 0° or 45°
2. **Video camera:** A high-resolution video camera attached to the eyepiece of the telescope acquires the image for projection on the monitor. The video image is transmitted *via* a cable to a video unit, where it is processed into either an analog or a digital form (Fig. 53.28).

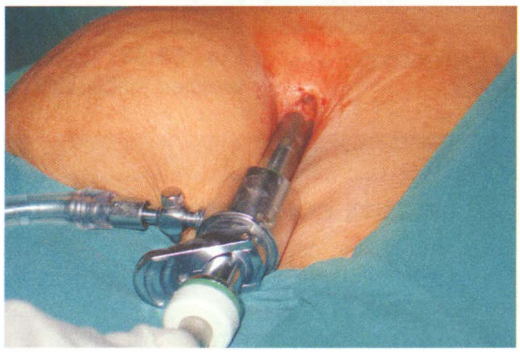


Fig. 53.28: Camera port (umbilical port)

- Analog is an electrical signal with a continuously varying wave or shift of intensity or frequency of voltage. Digital is a data signal with information represented by ones and zeros and is interpreted by a computer. These are the methods by which the picture is transmitted to the video monitor.
- **The camera and cable are designed so that they can be sterilised in glutaraldehyde**.

3. **Light sources:** High-intensity light is created with bulbs of mercury, halogen vapour or xenon. Since light is absorbed by blood, any procedure in which bleeding is encountered may require more light. The light is carried to the fibreoptic bundles of the laparoscope *via* a fibreoptic cable. The current systems create even brightness across the field.
4. **Insufflators:** An insufflator delivers gas from a high-pressure cylinder to the patient at a high rate with low and accurately controlled pressure (Fig. 53.29).
5. **Video monitors:** High-resolution video monitors are used to display the image. These monitors may be positioned optimally.

Fig. 53.29: Insufflator

Anaesthesia

- Usually done under general anaesthesia.
- Laparoscopic surgeon can influence cardiovascul performance by **reducing or removing the CO_2 pneum peritoneum**.
- **Insensible fluid losses are negligible**, and therefore, fluid administration should not exceed that necessary maintain circulating volume.
- Minimally invasive surgical procedures are often outpatie procedures. So, short-acting anaesthetic agents are preferab
- Since, the factors that require hospitalisation after lapar scopic procedures include the management of nausea, pa and urinary retention, the anaesthesiologist should minimi the use of agents that provoke these conditions and maximi the use of medications that prevent such problems.
- Critical to the anaesthesia management of these patients the **use of non-narcotic analgesics** (e.g. diclofenac) wh haemostasis allows it, and the **liberal use of antiemet** agents, such as **ondansetron and steroids.**

Procedure and principles

The unique feature of laparoscopic surgery is the need to li the abdominal wall from the abdominal organs by creatir pneumoperitoneum.

Gases used

Gases	Advantages	Disadvantages
Air	Historical importance	Poorly insoluble in blood so slower absorption More painful
CO_2	Inert	Rapidly absorbable Respiratory acidosis
N_2O	Inert Less painful Reduced intra-operative end tidal CO_2	Rapidly absorbable Danger of combustion Not safe in pregnancy

Laparoscopic access (Fig. 53.30)

- The requirements for laparoscopy are more involve because the creation of a pneumoperitoneum requires tha instruments of access (trocars) contain valves to maintai abdominal inflation.
- Two methods are used for establishing abdominal acces during laparoscopic procedures. The first, direct punctur laparoscopy, begins with the elevation of the relaxe abdominal wall with two towel clips or a well-placed hanc A small incision is made in the umbilicus, and a specialise spring-loaded (Veress) needle is placed in the abdomina cavity (Fig. 53.31). With the **Veress needle,** two distinc pops are felt as the surgeon passes the needle through th abdominal wall fascia and the peritoneum. The umbilicu usually is selected as the preferred point of access because in this location, the abdominal wall is quite thin, even i

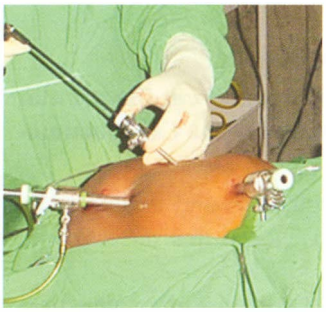


Fig. 53.30: 3 ports in place

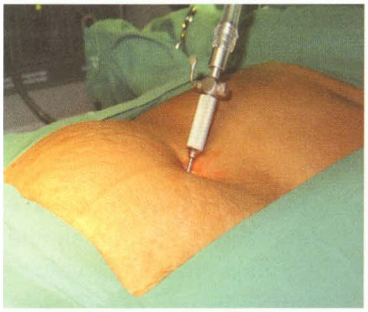


Fig. 53.31: Veress needle is used to create pneumoperitoneum

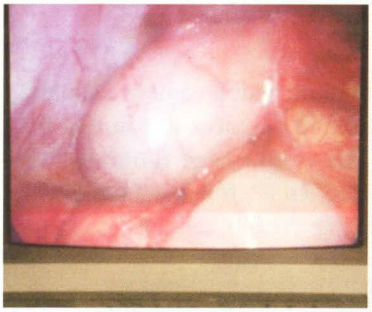


Fig. 53.32: Laparoscopic view of the gall bladder

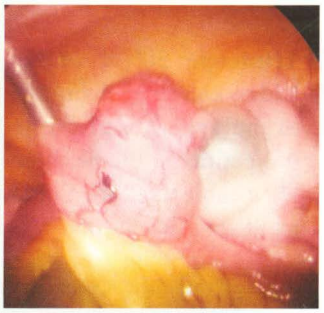


Fig. 53.33: Laparoscopic-assisted mobilisation of colon

obese patients. The abdomen is inflated with a pressure-limited insufflator. CO_2 gas is used usually with maximal pressures in the range of 14 to 15 mm Hg. During the process of insufflation, it is essential that the surgeon observe the pressure and flow readings on the monitor to confirm an intraperitoneal location of the Veress needle tip.

- Occasionally, the direct peritoneal access (Hasson) technique is advisable. With this technique, the surgeon makes a small incision just below the umbilicus and under direct vision locates the abdominal fascia.

Utility and scope

I. Basic

- Appendicectomy
- Cholecystectomy (Fig. 53.32)
- Hernia repair

II. Advanced

- Nissen fundoplication
- Heller's myotomy
- Gastrectomy
- Oesophagectomy
- Enteral access
- Bile duct exploration
- Colectomy (Fig. 53.33)
- Splenectomy
- Adrenalectomy
- Lymph node dissection
- Nephrectomy
- Robotics
- Stereo imaging
- Telemedicine

III. Laparoscopy-assisted procedures

- Hepatectomy
- Pancreatectomy
- Prostatectomy
- Hysterectomy

The physiologic effects of pneumoperitoneum

- The pneumoperitoneum has many effects that are only partially known despite years of study in humans and in animal models. There are effects resulting from the pressure within the abdomen and effects resulting from the composition of the gas used, generally carbon dioxide.
- The pressure within the abdomen from pneumoperitoneum decreases venous return by collapsing the intra-abdominal veins, especially in volume-depleted patients.
- This decrease in venous return may lead to decreased cardiac output.
- To compensate (Key Box 53.6), there is an elevation in the heart rate, which increases myocardial oxygen demand.
- High-risk cardiopulmonary patients cannot always meet the demand and may not tolerate a laparoscopic procedure. In volume-expanded healthy patients with full intra-abdominal capacitance vessels (veins), the increased intra-abdominal pressure actually may serve as a pump that increases right atrial filling pressure.
- Urine output often is diminished during laparoscopic procedures and usually is the result of diminished renal blood flow owing to the cardiovascular effects of pneumoperitoneum and direct pressure on the renal veins.
- In addition to direct effects, elevated intra-abdominal pressure results in release of antidiuretic hormone (ADH) by the pituitary, resulting in oliguria that may last up to 60 minutes after the pneumoperitoneum is released.

KEY BOX 53.6

ABSOLUTE CONTRAINDICATIONS

- Advanced generalised peritonitis
- Massive abdominal distension secondary to obstruction
- Irreducible hernia
- Uncorrected coagulopathy, hypovolaemic shock
- Inability of the patient to tolerate a formal laparotomy
- Surgeon's lack of experience in performing laparoscopic procedures

RELATIVE CONTRAINDICATIONS

• Previous surgery	Adhesions leading to visceral injury
• Ongoing intra-abdominal sepsis	Friable bowel prone to injury
• Bowel obstruction	Friable bowel prone to injury
• Morbid obesity	Difficult access, requirement for longer instruments.
• Pregnancy	Fetal distress, Injury to gravid uterus
• Aortic or iliac aneurysmal disease	Vascular injury
• Cardiopulmonary compromise	Raised intra-abdominal pressure may significantly reduce cardiac preload. CO_2 insufflation may result in CO_2 retention

Complications of laparoscopy

1. Injury to bowel/bladder
2. Injury to major vessels
3. CO_2 related complications

a. Hypercapnia: Hypercapnia and acidosis are seen with pneumoperitoneum and are likely due to the absorption of

carbon dioxide from the peritoneal cavity. Hypercapnia and acidosis that are difficult to control may follow, especially in elderly patients, those undergoing long operations and patients with pulmonary insufficiency.

b. Carbon dioxide embolus: The incidence of clinically significant CO_2 embolism is very low, although recent reports using more sensitive tests suggest that tiny bubbles of gas are present commonly in the right side of the heart during laparoscopic procedures. Clinically important CO_2 embolism may be noted by unexplained hypotension and hypoxia during the operation.

c. Capnothorax/pneumothorax: Capnothorax can be caused by carbon dioxide escaping into the chest through a defect in the diaphragm or tracking through fascial planes during dissection of the oesophageal hiatus. It can also be due to opening of pleuroperitoneal ducts most commonly seen on the right side.

HERNIA REPAIR : TAPP (Transabdominal Preperitoneal mesh repair)

KEY POINTS IN LAPAROSCOPIC INGUINAL ANATOMY

Space of Bogros

This 'preperitoneal space' is divided into two by the posterior lamina of the transversalis fascia. The posterior compartment of this space is called as the 'Space of Bogros (proper)', described by French anatomist Bogros in 1923. The anterior space has been termed as the 'Vascular Space'. Medially it is continuous with the space of Retzius.

Prevesical space of Retzius

The preperitoneal space that lies deep to the supravesical fossa and the medial umbilical fossa is the prevesical space of Retzius (described in 1858, by Swedish anatomist Retzius). Dissection of this space during a laparoscopic hernia repair is mandatory to enable proper mesh overlap of the hernial defect to aid in proper mesh placement/fixation.

Corona mortis' / crown of death / circle of death

- The pubic branch of the inferior epigastric artery courses in a vertical fashion inferiorly, crossing the Cooper's ligament and anastomosing with the obturator artery. In 25–30% of individuals (can be as high as 70-80%), the pubic branch is large and can replace the obturator artery.
- This large arterial branch is called aberrant obturator artery can partially encircle the neck of a hernia sac and be injured in a femoral hernia repair. It could also be injured while exposing the Cooper's ligament by freeing it of areolar adipose connective tissue.
- Because of this possibility an enlarged pubic branch of the Inferior epigastric artery has in the past been known as the – 'Corona Mortis'. The danger of injury in this area is more significant for obturator veins.

Triangle of doom

- It is a misnomer. It is not a triangle. It indicates an arc where it is dangerous to place staples or sutures durin laparoscopic hernia surgery.
- The "triangle of doom" is an inverted "V" shaped area wit its apex at the internal (deep) inguinal ring. The "triangl of doom" is bound laterally by the gonadal vessels, an medially by the vas deferens in the male, or the roun ligament of the uterus in the female.
- Within the boundaries of this area you can find the externa iliac artery and vein.
- Injury to these vessels can be catastrophic

Introduction

Novel method used for hernias wherein transabdominall (intraperitoneal) dissection is done through a laparoscope, an a mesh placed in the preperitoneal space.

Indication

Large indirect hernias and irreducible hernias.

Procedure

- **10 mm infraumbilical port** if used for the laparoscopi camera.
- **5 mm ports** are placed one on each side on pararectal poin at or above the level of umbilicus, so as to achieve adequat triangulation.
- Once ports are inserted, the hernial sac is recognised an the contents are reduced by pulling it transabdominall using a dissector (laparoscopic).
- Hernial sac is dissected in the preperitoneal plane afte incising at the upper part of the hernial sac opening.
- Once the sac is dissected and excised, a prolene mesh i placed in the preperitoneal space. It is fixed to the pubic bone using tacks. Peritoneum is closed with prolene sutures.

Complications

- Mesh displacement
- Intestinal obstruction if the mesh displaces into the peritoneum.
- Expensive
- Higher recurrence rates

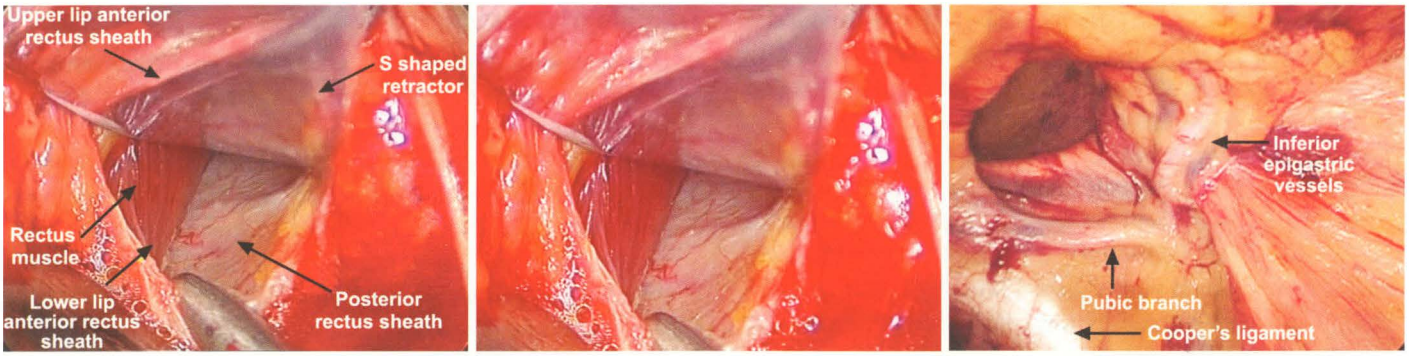
HERNIA REPAIR: TEP (totally extraperitoneal repair)

Indications

1. Recurrent hernia
2. Bilateral inguinal hernias
3. Indirect/direct/femoral hernias

Contraindications

1. Obstructed/strangulated hernias
2. Ascites
3. Bleeding disorders



Figs 53.34 to 53.36: Totally extraperitoneal repair (TEP) (*Courtesy:* Dr Praveen Bhatia, Consultant Surgeon and Medical Director, Bhatia Global Hospital and Endosurgery Institute, New Delhi)

This surgery has surpassed the TAPP procedure and is turning out to be a promising procedure for management of hernias.

Procedure (Figs 53.34 to 53.36)

- Subumbilical incision (10 mm) placed
- **Extraperitoneal space** is created by passing the scope between the rectus muscle and the posterior rectus sheath medial to the muscle bundle edge.
- Initial dissection is carried out using laparoscope itself and **inflation of CO_2.**
- 2 more 5 mm ports are placed in the midline **4 cm and 8 cm below the 1st port respectively**.
- **Dissection** is carried out medially till the pubic tubercle, iliopectinate ligament and laterally till the iliac vessels and inferior epigastric vessels. Once adequate space is dissected, the sac is reduced by pulling it down from the inguinal canal (reduction of sac) and a 15 × 15 cm mesh is placed and spread.
- Mesh may be sutured, left as it is or fixed with tackers.
- Both sides can be done together through the same ports.

Complications

1. Cord/vas injuries
2. Inadvertent opening of the sac/peritoneum and creation of pneumoperitoneum
3. Seroma formation
4. Infection

Advantages of TEP

1. Approach is totally extraperitoneal
2. Smaller incisions
3. No need for fixing mesh
4. Peritoneum is intact

SILS (LESS)

Introduction

- Single port access (SPA) surgery, also known as laparoendoscopic single-site surgery (LESS), single incision laparoscopic surgery (SILS) or single port incision less conventional equipment-utilising surgery (SPICES) or embryonic natural orifice transluminal endoscopic surgery (E-NOTES) is an advanced minimally invasive surgical procedure in which the surgeon operates almost exclusively through a single entry point, typically the patient's navel.
- SPA surgical procedures are like many laparoscopic surgeries in that the patient is under general anaesthesia; insufflated and laparoscopic visualisation is utilised.
- In laparoendoscopic single-site surgery (LESS), a single small incision is used at the entry point rather than four to five small incisions.
- All surgical instruments are placed through this small incision and also the incision site is located in the left abdomen or umbilicus. In general, SILS techniques take the same amount of time to do as traditional laparoscopic surgeries.
- However, SILS is recognised as to be a more complicated procedure because it involves manipulating three articulating instruments through one access port.
- Obesity, severe adhesions, or scarring from previous surgeries are a few cases, SILS may not be possible. Failure rates are high.

How SILS differs from traditional laparoscopic surgery?

- In single incision laparoscopic surgery, only one incision of around 1.5–2 cm is made just below the umbilicus to allow placement of three thin 5 mm port side by side parallel to each other.
- Port a, specially designed port is inserted into the abdomen; this port carries the telescope and laparoscopic instruments.
- Steps of surgical procedure are similar to the conventional laparoscopic surgery.
- As there is only one incision, pain is less as compared to traditional laparoscopic surgery and recovery is faster. The healed incision leaves practically no scar, thus making SILS cosmetically a superior option.
- In 5 to 10% patients, it may not be possible to complete the operation by SILS due to technical difficulties. One has to place one or two additional ports and completes the procedure in the traditional laparoscopic manner.

NATURAL ORIFICE TRANSLUMINAL ENDOSCOPIC SURGERY (NOTES)

Introduction

It means surgery performed endoscopically by initially passing the flexible endoscope through the body's natural orifices, like the mouth, anus, vagina, or urethra, to achieve access into areas that would not otherwise be accessible endoscopically, such as the abdomen and pelvis. Kalloo's did the first transgastric peritoneoscopy in 2004. In India, Dr GV Rao and Dr Nageshwar Reddy from Hyderabad, performed the first —NOTES in a patient who had appendicitis with extensive scars over the abdominal wall. The entry from abdomen was not possible. They performed transoral, transgastric appendicectomy.

Advantages

- Less invasive
- No abdominal incision
- Reduction in postoperative pain
- Wound infection, hernia formation and adhesions are very less.

Commonly performed NOTES are:

- Transgastric appendectomy
- Transvaginal cholecystectomy

Future upcoming technologies

- Magnetically anchored and guidance systems (MAGS) are designed to manoeuvre intra-abdominal instruments. They use the external handheld magnet.
- The fundus of the gall bladder can be retracted above the costal margin by coupling the interior aspect of an external magnet. The graspers are situated on the gall bladder with the help of endoscopic biopsy forceps.
- Magnets may become valuable, within the operating room.

VAAFT TECHNIQUE

Introduction

It is performed for the surgical treatment of complex anal fistulas and their recurrences. Key points are the exact localisation of the internal fistula opening under vision, the fistula treatment from inside, and the hermetic closure of the internal opening. No risk of faecal incontinence as no sphincter damage—one of the great advantages over the conventional treatment.

Materials

- Fistuloscope, a unipolar electrode connected to a high frequency unit, a fistula brush and a forceps.
- A semicircular or linear stapler and 0.5 ml of synthetic cyanoacrylate with a tiny catheter are used as well.
- The fistuloscope is equipped with an optical channel, a working channel and an irrigation channel. The working length adds up to 18 cm; the use of a handle reduces it to an effective length of 14 cm.
- The optimal patient positioning is the lithotomic position. Spinal anaesthesia is required.
- The fistuloscope is connected to the Karl Storz equipmen and to the washing solution bag (5000 cc glycine anc mannitol 1% solution).

The technique

It comprises a diagnostic phase and an operative phase.

The diagnostic phase

- The fistuloscope is inserted through the external fistula opening with the washing solution (glycine 1% anc mannitol 1%) already running. Thus, it provides clear view of the fistula pathway which is seen on the screen.
- With right index finger in the rectum, fistuloscope is guided slowly into the fistula.
- Complete relaxation of the surrounding tissue induced by the spinal anaesthesia helps in gentle up and down movements to advance the fistuloscope.
- The continuous flow of the glycine-mannitol solution allows for an optimal view of the fistula's inside up to the internal opening.
- At this stage, insert an anal retractor in order to localise the internal fistula opening by looking for the light of the telescope in the rectum or anal canal.
- When the fistuloscope exits through the internal opening the rectal mucosa clearly appears on the screen. At this point, two or three stitches are put, in two opposite points of the internal opening margin in order to isolate those points and not to lose them.

The operative phase

- First locate the internal opening. From the internal opening to external opening, the fistula wall and all granulation tissue are coagulated. Procedure is done slowly so that fistula is destroyed under vision using a unipolar electrode. All the necrotic material is removed. Abscess cavity is irrigated.
- Fistuloscope is removed at this stage. The assistant stretches the threads towards the internal rectal space or rather the anal canal using a straight forceps in order to lift the internal fistula opening at least 2 cm into the shape of a volcano.
- Subsequently, stitch is inserted at the volcano's base and complete the mechanical cutting and suturing. By using a linear stapler. The hermetic closure of the internal fistula opening can also be accomplished. This also depends on the internal opening position. Using a semicircular stapler, the suture will be horizontal. Using a linear stapler, the suture will be vertical.
- Last step is insertion of 0.5 ml of synthetic cyanoacrylate after the suture/staple line via the fistula pathway to further reinforce the suture. It helps in perfect closure of the fistula opening.
- This procedure assures a perfect excision and a hermetic closure of the internal fistula opening, excluding the risk of stool passage. Since the suture is situated tangential to the sphincter, the postoperative pain is low even if the suture falls both in the anal canal and the rectum.

Conclusion

- The advantages of the VAAFT technique are: No surgical wounds on the buttocks or in the perianal region, there is complete certainty in the localization of the internal fistula opening, and the fistula can be completely destroyed from the inside.
- Since operations are done from inside, no damage is caused to the anal sphincters. The risk of postoperative faecal incontinence is excluded.

HIGH FREQUENCY (HF) ELECTROSURGERY

Principle involves passage of electric current through tissue by means of potential difference (voltage). The resultant flow of electrons excites the tissue molecules, notably water, creating heat energy which causes water evaporation and tissue coagulation. HF electrosurgery can be monopolar or bipolar. Here, the current escapes from electrode tip into the receptive tissue and exits through the grounding pad.

Unmodulated continuous sine wave in voltage range 200–500 mV is used for **electrocutting** (Fig. 53.37).

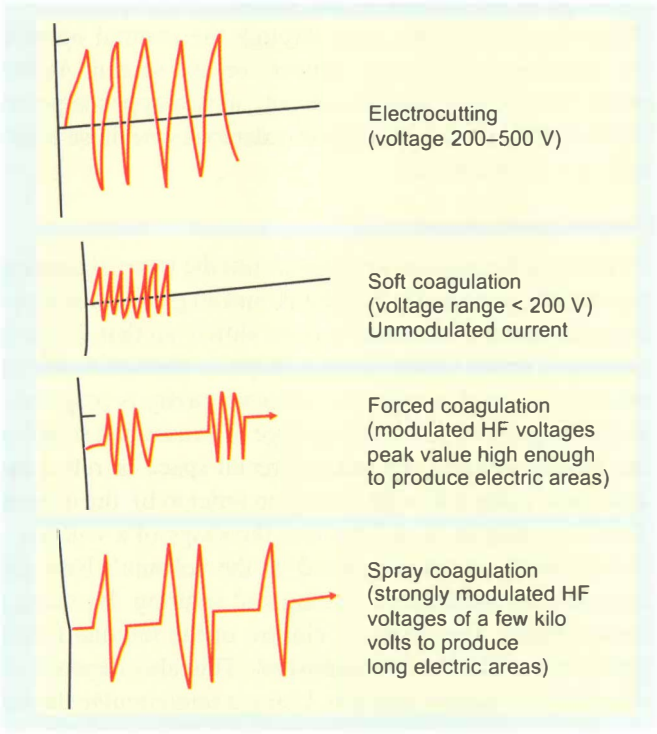


Fig. 53.37: Electrocautery

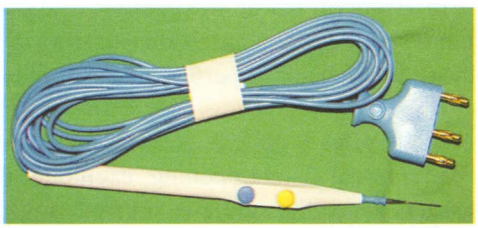


Fig. 53.38: Cautery cord: Yellow button is for cutting current and blue button is for coagulation

Uses of electrocautery

1. To achieve haemostasis
2. Removal of skin tags
3. Treating very small, early basal cell carcinoma
4. Removal of erosions of cervix
5. Removal of condylomata, cutaneous acanthoma, warts, etc.

Bipolar electrocautery

Heat energy is concentrated between two electrodes and does not dissipate throughout the tissue. Hence,

- Small volume of tissue is injured
- Less risk of burning injury
- Safe with pacemakers
- Excellent for obtaining haemostasis in areas that may be in close proximity to delicate structures, e.g. head and neck surgery.

PEARLS OF WISDOM

Effective in wet fields, uses coagulation current only.

Monopolar electrocautery

Heat energy and thus tissue injury can extend for some distance away from the point of contact. Hence, it should not be used with

- Direct contact with a hollow viscus as this may lead to perforation.
- Close proximity to a major blood vessel as it may cause vessel wall injury.

PEARLS OF WISDOM

Not effective in wet fields, uses both cutting and coagulation current. Therefore, dissection is possible.

CRYOSURGERY

Definition

The application of a freezing probe to the living tissues is termed **cryosurgery.**

Mechanism of tissue destruction

- Freezing produces ice crystal formation within cell wall resulting in cell membrane rupture and death.
- Also as the crystals grow, water is removed from neighbouring cells leading to an increase in the concentration of electrolytes. These soon reach toxic levels, initiating cell death by osmotic shock.

Methodology and instrument used

- **N_2O system** can achieve a probe tissue temperature of around –90°C. It works with the principle of rapid expansion of a gas producing refrigeration (Joule-Thompson effect).
- Liquid nitrogen (N_2) system can achieve a probe temperature of around –190°C.

Uses

1. Haemorrhoidectomy
2. Cutaneous lesions---excision or biopsy

LASERS IN SURGERY

- **L**ight **A**mplification by **S**imulated **E**mission of **R**adiation
- Molecules which are placed in a compact are activated when power is passed through.
- As a result of this they move in different directions, they hit each other, releasing energy. This energy is used as laser to the area whenever required.

Types

1. Argon laser
2. Neodymium: Yttrium-aluminium-garnet laser (Nd:YAG laser)
3. CO_2 laser
4. Neon laser

Advantages and disadvantages

- Most important advantage is a bloodless field—specially useful in head and neck surgeries and ENT surgeries.
- It is quick and there is less tissue trauma
- Expensive

Precaution

To avoid injuries to normal tissues, all reflecting instruments should be avoided so that the laser does not get reflected.

PEARLS OF WISDOM

All theatre personnel should wear special protective goggles.

Clinical applications

1. Vascular malformation of the GIT
2. Endoscopic laser for advanced carcinoma oesophagus to relieve obstruction and dysphagia.
3. Obstructed colorectal cancer
4. **Liver resections:** Nd:YAG laser combined with CUSA can be used for liver resections.
5. CO_2 laser and Nd:YAG laser can be used for haemorrhoidectomy.

STAPLERS IN SURGERY

Principle

They are used for apposition of tissues.

Types

1. Cutaneous staplers
- Used after thyroidectomy. It is quick and gives clean apposition.
- Needs a special instrument for removal.

2. Linear staplers
- Used to close the bowel partially or completely.

3. Circular staplers
- Are also called EEA stapler: End-to-end anastomosis.
- Uses in surgery:
  a. After low or high anterior resection done for carcinoma rectum
  b. After oesophagogastrectomy
  c. Any other intestinal resection

4. GIA stapler (Fig. 53.39)
- Gastrointestinal anastomosis stapler: Used for side-to-side anastomosis.

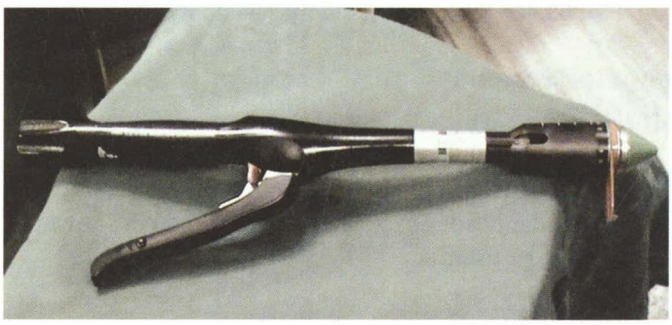


Fig. 53.39: GIA stapler

5. Endostapler
- With the increasing use of laparoscopy surgeries for facilitating a quick and safe anastomosis, endostaplers are used for intestinal anastomosis.
- Endovascular staplers are used to ligate vascular pedicles Examples: **Renal pedicles** during laparoscopic nephrectomy, **adrenal veins** during laparoscopic adrenalectomy.

Advantages of staplers

- Saves operating time
- The low rectal and oesophageal anastomosis have higher incidence of leakage rates. However, it can be decreased by using staplers.

Disadvantages

- Expensive
- Improper apposition results in leakage

Parts of the stapler

1. Handle
2. Shaft
3. Head, detachable anvil + a staple cartridge.
   The staples (approximately 15 in number) are present in the cartridge. The cartridge also has a circular knife.

PEARLS OF WISDOM

The doughnuts (rings of excised tissue) should be complete after the stapled anastomosis. Incomplete doughnut means incomplete wound closure (Fig. 53.40).

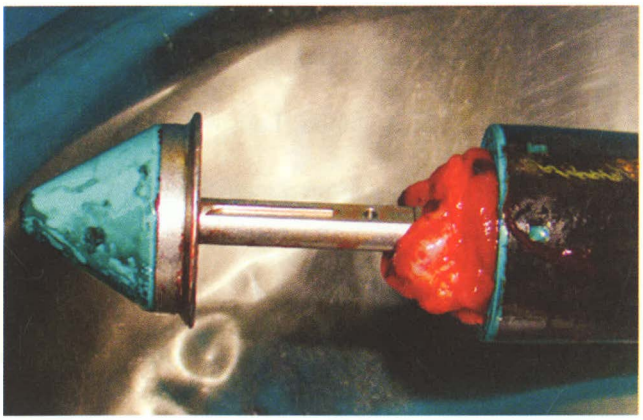


Fig. 53.40: Doughnut of stapler haemorrhoidopexy

Contraindications

1. If the tissues which have to be approximated are under tension, they should not be stapled.
2. Different lumen diameters should not be stapled end-to-end
3. If the circular head is of greater diameter than the lumen, it should not be used.

MISCELLANEOUS

WHEN TO DO PROPHYLACTIC SURGERY?

- Prophylactic **bilateral mastectomy** in BRCA 1 and BRCA 2 patients
- Prophylactic **total colectomy and ileoanal pouch** in familial polyposis coli patients
- Prophylactic **total thyroidectomy** in familial medullary carcinoma thyroid patients
- Prophylactic **cholecystectomy** in Pima Indians
- Prophylactic **vagal sparing transhiatal oesophagectomy** (THE) for severe dysplasia
- Prophylactic **gastrectomy-E-cadherin** mutation

PLEASE READ THESE INSTRUCTIONS

I have added colectomy, cholecystectomy, GJ vagotomy, amputations, laparoscopic surgeries including hernia as per wishes of many students who have read 3rd edition. Students are requested to confirm the operations list which will be asked in the examination with their teachers in their respective medical colleges and be prepared for exams. You should realise that what operation an undergraduate student is expected to know in more detail is not stated clearly in the syllabus. Nevertheless, you do not lose anything trying to understand more operations. Rather, it may help you in your postgraduate entrance examinations.

WHAT IS NEW IN THIS CHAPTER? / RECENT ADVANCES

- All topics have been updated
- Cholecystectomy, GJ vagotomy, colectomy and laparoscopic surgery have been included
- Excision of swellings has been included
- Laparoscopic surgery have been upgraded
- SILS, NOTES, VAAFT have been added

- ***The last four chapters are important for viva voce examination in general surgery. The questions given in these chapters are most commonly asked. This does not mean, however, that they are the only questions asked. As the subject is vast, the number of questions that can be asked can be unlimited. The purpose of viva voce section is to see how much the student knows as well as the depth and understanding of the subject. Our best wishes to you once again. Enjoy reading Manipal Manual of Surgery, 4th edition—Authors***

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